Posterior cortical atrophy

This Help Sheet has been developed to assist your understanding of posterior cortical atrophy and its symptoms, diagnosis, progression and treatment.

What is posterior cortical atrophy?

Posterior cortical atrophy (PCA) is a progressive degenerative condition involving the loss and dysfunction of brain cells particularly at the back (posterior) of the brain. Brain tissue in the outer layer (cortical) shrinks (atrophy) as cells are lost. In the vast majority of cases, this loss of brain cells is associated with the same pathological brain changes seen in typical Alzheimer's disease, namely amyloid plaques and neurofibrillary tangles. In other words, PCA is most usually considered to be an unusual or atypical variant of Alzheimer's disease.

Differences to typical Alzheimer's disease

Despite being caused by the same disease process, the effects of PCA and typical Alzheimer's disease on the behaviour, thinking and skills of individuals with each condition can be very different. Typical Alzheimer's disease is most commonly associated with deterioration in memory, language, perception and other skills and abilities. By contrast, individuals with PCA tend to have well preserved memory and language but instead show a progressive, dramatic and relatively selective decline in visual processing and/or literacy skills such as spelling, writing and arithmetic.

The reason for this marked distinction between PCA and typical Alzheimer's disease is that the distribution of the disease in the brain is different. In typical Alzheimer's disease, disease-related changes are evident across most brain areas, whereas in PCA the changes are restricted to or focussed on the back of the brain and so initially only the skills which those brain areas normally support are affected.

Symptoms of posterior cortical atrophy

Difficulties with skills such as literacy, numeracy, and the ability to make skilled movements may be among the first symptoms noticed by individuals with PCA. Such difficulties may be experienced in the following ways:

- Problems recalling the exact spelling of words
- Difficulties with handwriting or typing
- Difficulties with remembering the shape or name of particular letters or numbers
- Slowness and difficulty with mental arithmetic
- Problems dealing with money and small change
- Awkwardness making gestures (for example, waving, thumbs up)
- Difficulties with using particular tools, kitchenware or implements (for example, cutlery, scissors, glasses)
- Problems with dressing and clothing (partly related to difficulties with visual perception)

Another common problem noticed by individuals with PCA is with visual processing. Quite understandably, this leads many people to consult an eye specialist, but in fact the visual problems experienced are not related to the eye. Rather, the problems stem from the difficulty the affected brain has with interpreting the information sent to it by the healthy eyes. The precise nature of the visual problems experienced may vary widely but often include difficulty with some or all of the following:

- Recognising objects in pictures (particularly if taken from obscure angles or incomplete)
- Recognising faces (for example, television characters, friends, relatives)
- Appreciating the spatial location of objects (for example, missing when reaching out to pick something up)
- Judging speed or distance (for example, when driving a car or walking down stairs)
- Perceiving movement among things which are stationary
- Following text when reading (for example, missing some lines of text)
- Reading particular words (for example, finding that letters appear to move around)
- Reading certain types of text (for example, large print such as newspaper headlines)
- Experiencing objects as having an unusual colour
- Experiencing increased sensitivity to bright light or shiny surfaces
- Seeing clearly (for example, experiencing double vision, or feeling that one's eyes are jerking around)

This help sheet is funded by the Australian Government

PCA can affect people in different ways initially. In some instances, the disease affects both sides of the brain equally, leading to a combination of many of the symptoms described above. For other people, the disease affects one particular brain area earlier or more significantly. As a result, problems with spelling and writing, for example, might be the first sign of the condition with vision relatively unaffected, whereas for others, difficulties in seeing where objects are might be the initial symptom.

Diagnosis of posterior cortical atrophy

PCA tends to affect people at an earlier age than typical Alzheimer's disease, with individuals often being in their mid-fifties or early sixties at the time of their first symptoms. However, it can also affect older people. The first signs are often subtle symptoms which may be difficult for the person experiencing them to explain. Individuals with early visual complaints are typically referred to eye specialists before referral to a neurologist. However, other problems with spelling, numeracy, and learned motor skills have also been reported. Even once an appropriate referral has been made, it may take some time before the diagnosis is made formally.

There is no diagnostic test for PCA. However, a number of tests may help to exclude potentially treatable causes such as infection or brain tumours. These may include some or all of the following:

- Specialised visual tests (by eye specialists)
- A full neuropsychological assessment (tests of thinking and reasoning abilities)
- Laboratory blood tests
- Brain imaging
- Lumbar puncture (examination of the fluid circulating around the brain and spinal cord)
- Other medical tests

The shrinkage of the back part of the brain as a result of brain cell loss may be visible on a brain scan. However, it may be difficult to diagnose, and a definitive diagnosis can only be made post-mortem once the brain tissue is examined by a pathologist.

The underlying cause of PCA in the majority of cases is Alzheimer's disease. However, other conditions can also result in similar initial symptoms. These include Lewy body disease, corticobasal degeneration and Creutzfeldt-Jakob disease. The exact prevalence of PCA in the population is currently unknown.

Progression of posterior cortical atrophy

As PCA progresses, word finding, day-to-day memory and general cognitive functions may become affected. In the later stages of the disease, people with PCA may develop jerking movements of their limbs and occasionally seizures. PCA causes progressive and irreversible decline in a person's skills and abilities over the years following diagnosis, and is a terminal disease.

Treatment of posterior cortical atrophy

Although there is no medication available to treat PCA specifically, patients may find some of the medications available for the treatment of typical Alzheimer's disease helpful. However, these medications are only designed to treat the symptoms of the disease. They may slow the disease progression but they are not a cure.

Although individuals with PCA often show relatively preserved insight, the cognitive problems associated with the condition often lead to a loss of independence, cause activities of daily living to be compromised, and inhibit the enjoyment of previous hobbies. Such problems can lead to depression, irritability, frustration and a loss of self-confidence. Individuals with PCA who are experiencing low mood may benefit from psychological therapies or antidepressant medication.

Practical visual aids designed to assist people with visual impairment may be of use to people with PCA. These include devices such as talking clocks or watches, mobile phones with simplified displays, cooking aids such as sensors that beep when a cup is almost full, and audio books.

Support for people with posterior cortical atrophy

Vision Australia provides advice and products that may be of help to people with PCA. Visit **visionaustralia.org**

The Royal Society of The Blind and Canberra Blind Society provide a full range of services for people who are blind or vision impaired. Visit **rsb.org.au**

FURTHER INFORMATION

DementiaAustralia offers support, information, education and counselling. Contact the National Dementia Helpline on **1800 100 500**, or visit our website at **dementia.org.au**

Interpreter For language assistance phone the Translating and Interpreting Service on **131 450**

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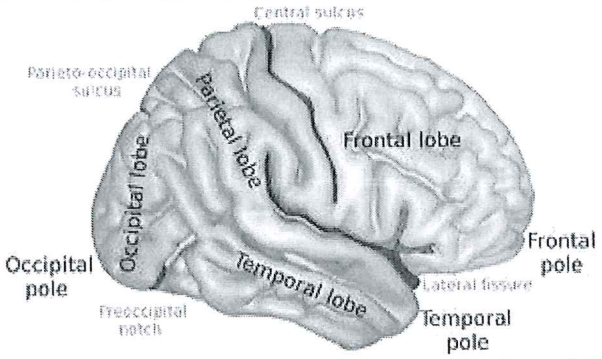
Posterior cortical atrophy

Posterior cortical atrophy (**PCA**), also called **Benson's syndrome**, is a form of dementia which is usually considered an atypical variant of Alzheimer's disease (AD).[1][2] The disease causes atrophy of the posterior part of the cerebral cortex, resulting in the progressive disruption of complex visual processing.[3] PCA was first described by D. Frank Benson in 1988.[4][5]

PCA usually affects people at an earlier age than typical cases of Alzheimer's disease, with initial symptoms often experienced in people in their mid-fifties or early sixties.[3] This was the case with writer Terry Pratchett (1948-2015), who went public in 2007 about being diagnosed with PCA.[6] In rare cases, PCA can be caused by dementia with Lewy bodies and Creutzfeldt–Jakob disease.[5][3]

Posterior cortical atrophy

Other names	Biparietal Alzheimer disease



Lobes of the human brain

Specialty	Neurology

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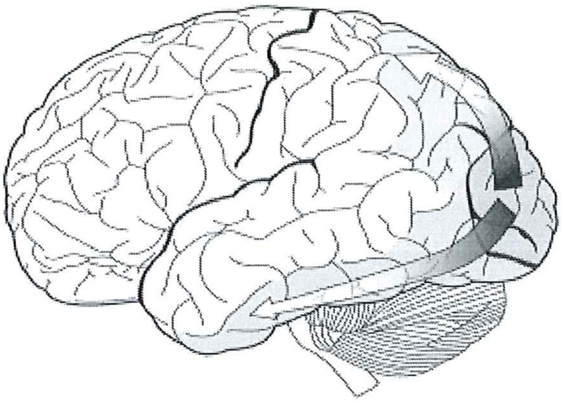
Symptoms

The main symptom resulting from PCA is a decrease in visuospatial and visuoperceptual capabilities.[7] Because the posterior region of the brain is home to the occipital lobe, which is responsible for visual processing, visual functions are impaired in PCA patients. The atrophy is progressive; early symptoms include difficulty reading, blurred vision, light sensitivity, issues with depth perception, and trouble navigating through space.[8][9] Additional symptoms include apraxia, a disorder of movement planning, alexia, an impaired ability to read, and visual agnosia, an object recognition disorder.[10] Damage to the ventral, or "what" stream, of the visual system, located in the temporal lobe, leads to the symptoms related to general vision and object recognition deficits; damage to the dorsal, or "where/how" stream, located in the parietal lobe, leads to PCA symptoms related to impaired movements in response to visual stimuli, such as navigation and apraxia.[10][9]

As neurodegeneration spreads, more severe symptoms emerge, including the inability to recognize familiar people and objects, trouble navigating familiar places, and sometimes visual hallucinations.[7][8] In addition, patients may experience difficulty making guiding movements towards objects, and may experience a decline in literacy skills including reading, writing, and spelling.[8][11][12] Furthermore, if neural death spreads into other anterior cortical regions, symptoms similar to Alzheimer's disease, such as memory loss, may result.[8][11] PCA patients with significant atrophy in one hemisphere of the brain may experience hemispatial neglect, the inability to see stimuli on one half of the visual field.[9] Anxiety and depression are also common in PCA patients.[13]



The dorsal stream (green) runs through the parietal lobe, and ventral stream (purple) runs through the temporal lobe. Both streams originate in the occipital lobe (blue) located posteriorly.

Connection to Alzheimer's disease

Studies have shown that PCA may be a variant of Alzheimer's disease (AD), with an emphasis on visual deficits.[1][10] Although in primarily different, but sometimes overlapping, brain regions, both involve progressive neural degeneration, as shown by the loss of neurons and synapses, and the presence of neurofibrillary tangles and senile plaques in affected brain regions; this eventually leads to dementia in both diseases.[14][15] PCA patients have more cortical damage and gray matter (cell body) loss in posterior regions, especially in the occipital, parietal, and temporal lobes, whereas Alzheimer's patients typically experience more damage in the prefrontal cortex and hippocampus.[11][14][16] PCA tends to impair working memory and anterograde memory, while leaving episodic memory intact, whereas AD patients typically have damaged episodic memory, suggesting some differences still lie in the primary areas of cortical damage.[8][14]

Over time, however, atrophy in PCA patients may spread to regions commonly damaged in AD patients, leading to common AD symptoms such as deficits in memory, language, learning, and cognition.[10][11][14][15] Although PCA has an earlier onset, many PCA patients have also been diagnosed with Alzheimer's, suggesting that the degeneration has simply migrated anteriorly to other cortical brain regions.[7][10]

There is no standard definition of PCA and no established diagnostic criteria, so it is not possible to know how many people have the condition. Some studies have found that about 5 percent of people diagnosed with Alzheimer's disease have PCA. However, because PCA often goes unrecognized, the true percentage may be as high as 15 percent. Researchers and physicians are working to establish a standard definition and diagnostic criteria for PCA.[17]

PCA may also be correlated with the diseases of Lewy body, Creutzfeldt–Jakob disease, Bálint's syndrome, and Gerstmann syndrome.[8][9][18] In addition, PCA may result in part from mutations in the presenilin 1 gene (PSEN1).[9]

Diagnosis

At this time the cause of PCA is unknown; similarly, there are no fully accepted diagnostic criteria for the disease.[1][9] This is partially due to the gradual onset of PCA symptoms, the variety of symptoms, the rare nature of the disease and younger age of patients (initial symptoms appear in patients of 50–60 years old).[19] In 2012, the first international conference on PCA was held in Vancouver, Canada. Continued research and testing will hopefully result in accepted and standardized criteria for diagnosis.[9]

PCA patients are often initially misdiagnosed with an anxiety disorder or depression. Some believe that patients may experience depression or anxiety due to their awareness of their symptoms, such as decrease in their vision capabilities, yet they are unable to control this decline in their vision or the progressive nature of the disease. The early visual impairments of a PCA patient have often led to an incorrect referral to an ophthalmologist, which can result in unnecessary cataract surgery.[19]

Due to the lack of biological marks of PCA, neuropsychological examinations should be used for diagnosis.[20] Neuroimaging can also assist in diagnosis of PCA.[19] The common tools used for neuroimaging of both PCA and AD patients are magnetic resonance imaging (MRI's), a popular form of medical imaging that uses magnetic fields and radio waves, as well as single-photon emission computed tomography, an imaging form that uses gamma rays, and positron emission tomography, another imaging tool that creates 3D images with a pair of gamma rays and a tracer.[21] Images of PCA patient's brains are often compared to AD patient images to assist diagnosis. Due to the early onset of PCA in comparison to AD, images taken at the early stages of the disease will vary from brain images of AD patients. At this early stage PCA patients will show brain atrophy more centrally located in the right posterior lobe and occipital gyrus, while AD brain images show the majority of atrophy in the medial temporal cortex. This variation within the images will assist in early diagnosis of PCA; however, as the years go on the images will become increasingly similar, due to the majority of PCA patients also having AD later in life because of continued brain atrophy.[9][22] A key aspect found through brain imaging of PCA patients is a loss of grey matter (collections of neuronal cell bodies) in the posterior and occipital temporal cortices within the right hemisphere.[23]

For some PCA patients, neuroimaging may not result with a clear diagnosis; therefore, careful observation of the patient in relation to PCA symptoms can also assist in the diagnosis of the patient.[19] The variation and lack of organized clinical testing has led to continued difficulties and delays in the diagnosis of PCA in patients.[9]

Treatment

Specific and accepted scientific treatment for PCA has yet to be discovered; this may be due to the rarity and variations of the disease.[9][24] At times PCA patients are treated with prescriptions originally created for treatment of AD such as, cholinesterase inhibitors, donepezil, rivastigmine and galantamine, and memantine.[9] Antidepressant drugs have also provided some positive effects.[19]

Patients may find success with non-prescription treatments such as psychological treatments. PCA patients may find assistance in meeting with an occupational therapist or sensory team for aid in adapting to the PCA symptoms, especially for visual changes.[9][19] People with PCA and their caregivers are likely to have different needs to more typical cases of Alzheimer's disease, and may benefit from specialized support groups such as the PCA Support Group (http://www.raredementiasupport.org/pca/) based at University College London, or other groups for young people with dementia. No study to date has been definitive to provide accepted conclusive analysis on treatment options.[19]

References

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University of California San Francisco

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Posterior Cortical Atrophy

Posterior cortical atrophy (PCA), also called Benson's syndrome, is a rare, visual variant of Alzheimer's disease. It affects areas in the back of the brain responsible for spatial perception, complex visual processing, spelling and calculation.

What Causes PCA?

In the vast majority of PCA cases, the underlying cause is Alzheimer's disease, and the brain tissue at autopsy shows an abnormal accumulation of the proteins amyloid and tau that form the plaques and tangles seen in Alzheimer's disease. Although PCA is almost always caused by Alzheimer's disease, it can result from other diseases including dementia with Lewy bodies and Creutzfeldt-Jakob disease. PCA is thought to affect less than 5% of people with Alzheimer's disease, although epidemiological studies are lacking and PCA has been under-recognized in the past.

How is Age Related to PCA?

People with PCA may often go to see an eye doctor first, thinking that their difficulties are due to a problem with their eyes and that they may need new glasses. Visual impairment commonly develops as people get older, and in most cases, a decline in vision is due to this natural aging process. In people with PCA, the visual problems are not due to problems with their eyes. Rather, the shrinking brain can no longer interpret and process the information received from the person's healthy eyes.

What Happens in PCA?

Early symptoms of posterior cortical atrophy include blurred vision, difficulties reading (particularly following the lines of text while reading) and writing with non-visual aspects of language preserved, problems with depth perception, increased sensitivity to bright light or shiny surfaces, double vision and difficulty seeing clearly in low light conditions. The patient may have trouble accurately reaching out to pick up an object. As the disorder progresses, other symptoms evolve such as getting lost while driving or walking in familiar places, misrecognition of familiar faces and objects, and rarely visual hallucinations. Calculation skills and the ability to make coordinated movements are affected in some cases.

While there is no definitive test for PCA, neuropsychological evaluation of cognitive skills, blood tests, brain scans and a neurological exam may help to exclude potentially treatable causes such as infection, inflammation or brain tumor.

PCA tends to affect people at an earlier age than typical Alzheimer's disease, with individuals often being in their mid-fifties or early sixties when they experience the initial symptoms. As the disease progresses, word finding, day-to-day memory and general cognitive functions may become affected, and people develop the symptoms of typical Alzheimer's disease. In the later stages of the disease, people may experience jerking movements of their limbs and even seizures. Some people live approximately the same length of time as individuals with typical Alzheimer's disease (on average 10–12 years following the onset of symptoms) while others live with the condition for longer.

Are There Medicines to Treat PCA?

Although no cure for posterior cortical atrophy exists, several medications, as well as many non-pharmaceutical approaches, can potentially improve daily functioning and quality of life. Patients with posterior cortical atrophy can often benefit from physical and occupational therapy.

Cholinesterase inhibitors approved for Alzheimer's disease, like donepezil (Aricept®), rivastigmine (Exelon®) and galantamine (Razadyne®), can help the symptoms of PCA by boosting the function of brain cells to compensate for damage caused by Alzheimer's disease.

Patients experiencing depression, irritability, frustration and a loss of self-confidence may benefit from antidepressant medication.

Resources

- Unraveling Posterior Cortical Atrophy: Not Seeing Things That Are There, clinical story by Jamie Talan, Atlantic Fellow at GBHI
- Alzheimer's Disease Education and Referral (ADEAR) Center
    - Alzheimer's Disease Fact Sheet
    - Find an Alzheimer's Disease Center (ADC)
- The Alzheimer's Association, Northern California and Northern Nevada chapter
- Home Safety Tips & Recommendations for People with Visual Dysfunction
- Family Caregiver Alliance
- Rare Dementia Support: Posterior Cortical Atrophy Support
- Posterior Cortical Atrophy Support Group on Facebook
- International Consensus on the First Diagnostic Guidelines for Posterior Cortical Atrophy
- Dementia Resources for Health Professionals

Participate in Research

- ADRC: New Approaches to Dementia Heterogeneity
- Alzheimer's Disease Neuroimaging Initiative (ADNI)

Posterior Cortical Atrophy - a rare form of dementia

Posterior cortical atrophy (PCA) is an atypical form of dementia primarily affecting sight rather than memory. It is a progressive condition where the degeneration and loss of brain cells is found mostly in the back (posterior) part of the brain.

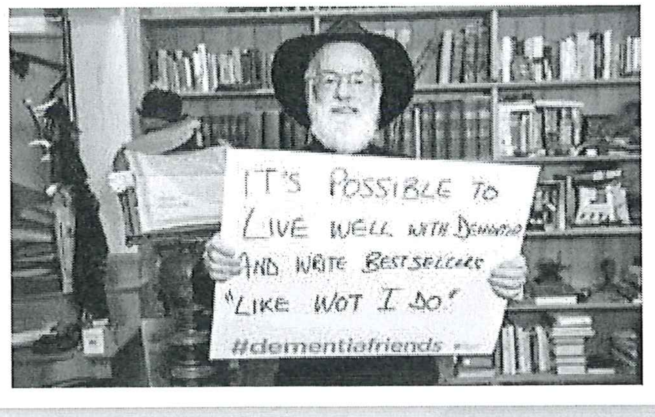
In most cases, the same pathological changes that are seen in Alzheimer's disease (loss of brain cells associated with the presence of amyloid plaques and neurofibrillary tangles) are also found in the brains of people with PCA. PCA is therefore considered to be an unusual variant of Alzheimer's disease.

Although the same disease process occurs in both Alzheimer's disease (AD) and PCA, the effects of these two conditions are markedly different. In AD, deterioration in memory, language, perception and insight are typically seen. In PCA, however, memory, insight and language tend to be preserved (at least in the early stages) but there is a specific decline in vision and literacy skills such as spelling, writing and arithmetic. The reason for this difference is that AD-related changes occur across most brain areas whereas PCA-related changes are restricted to the rear of the brain where visual processing, literacy, numeracy and motor planning and execution (praxis) are focussed.

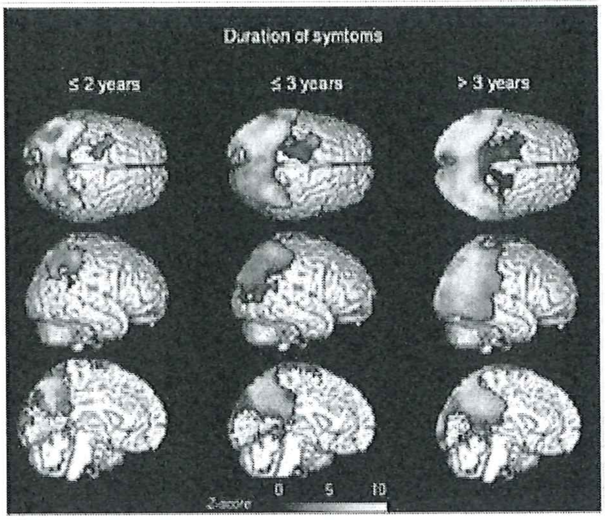
Initial symptoms of PCA often include difficulties with driving (including judging distances and speed), fine spatial judgements (e.g. missing when reaching out to pick something up), recognition of what or where objects are (e.g. difficulty recognising common household items in a catalogue) and deterioration in reading, writing, spelling and counting abilities.

Because the most common problems noticed by individuals with PCA are with vision, people with PCA often first consult their optometrist or ophthalmologist. However, the difficulties being experienced are not related to the eyes but to the back part of the brain which cannot correctly process the information sent to it by the eyes. PCA is therefore not a problem of eyesight but of brain function.

PCA often occurs in younger people, with the early symptoms often presenting in the 50s and 60s.



Sir Terence David John "Terry" Pratchett, OBE, English author of fantasy novels, diagnosed with PCA in 2007 (Source: Dementia Vision, http://livingwelldementia.org/2014/05/16/terry-pratchett-norman-mcnamara-shibley-rahman-living-well-with-dementia/)



(Source: Dementia Vision, http://livingwelldementia.org/2014/05/31/meeting-other-people-living-with-posterior-cortical-atrophy/)

Because PCA often occurs in younger people and has an unusual presentation, diagnosis of PCA is often slow and frustrating for the person affected. Early symptoms are often attributed to other causes such as impaired eyesight, anxiety, depression, menopause or even malingering and typically time to diagnosis of PCA from the onset of symptoms is one to three years.

In the moderate stages of PCA, common problems include: difficulty navigating between rooms or locating particular places such as the bathroom; difficulty recognising even very familiar faces; movement problems such as being able to walk but not sit down; difficulty with spatial commands or actions such as "turn around" or "shuffle forward"; and difficulties with handwriting, mental arithmetic and spelling.

In the later stages of the condition, word-finding, day-to-day memory and general cognitive functions can become affected. Jerking movements of the limbs (myoclonus) and occasionally seizures may also occur at this late stage. Further information on the symptoms of PCA can be found in "The Stages of PCA" available at https://www.ucl.ac.uk/drc/pcasupport/stages-of-pca

While there is no medication available to treat PCA specifically, there is some evidence to support the use of the medications available to treat AD, namely donepezil (Aricept), rivastigmine (Exelon) and galantamine (Reminyl). Although these medications are not a cure for PCA, they are designed to treat the symptoms of the condition.

Because people with PCA often show relatively preserved insight into their condition, peer support groups, group, couple and individual psychological therapies and use of practical aids such as talking clocks/watches, mobile phones with simplified displays, talking books and voice recognition software can be of benefit.

References can be obtained by contacting 1800 639 331.

⚠ Coronavirus information for **Feinberg (https://www.feinberg.northwestern.edu/sites/covid-19/)**.

Symptoms & Causes of Posterior Cortical Atrophy

Symptoms vary from individual to individual and will change as the disease progresses. Early on patients may experience subtle visual misperceptions — when the person sees one thing as something else — which, naturally, is thought to be a problem with their eyes or their glasses. However this is not the case.

An individual with Posterior Cortical Atrophy (PCA) will notice these changes with vision develop gradually over months to years often leading to:

- Inability to recognize common objects
- Inability to judge spatial relationships or reach for objects accurately
- Difficulty using common items, like a phone.
- Difficulty finding things in plain sight
- Inability to read or write
- Inability to dress
- Other abilities controlled by the back parts of the brain, such as math abilities or discerning left from right, may be affected

In the later stages of the disease patients are functionally blind, which means a person loses their sight. Though initially spared, other aspects of cognition such as memory, executive function, and language may eventually be affected.

PCA is most commonly caused by Alzheimer's Dementia. Other neurodegenerative disorders like Lewy Body Dementia, Corticobasal Degeneration, and prion disease (Creutzfeldt-Jakob Disease) may rarely lead to this syndrome.

More in Posterior Cortical Atrophy

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Diagnosis of Posterior Cortical Atrophy

The Neurobehavior and Memory Clinic at Northwestern Medicine **(https://www.nm.org/conditions-and-care-areas/neurosciences/neurobehavior-and-memory-clinic)** is affiliated with our center and offers clinical consultations to patients, families and providers, including diagnostic evaluations, second opinions and supportive services.

After cognitive testing with a doctor or neuropsychologist has determined there is a problem, an MRI is often the next step in diagnosis of Posterior Cortical Atrophy (PCA). An MRI rules out strokes, tumors, and other structural problems while looking for characteristic "atrophy" or shrinking in the back parts of the brain. This atrophy suggests a neurodegenerative process may be ongoing. Typically blood work is done to rule out thyroid problems, infections, or vitamin deficiencies that rarely contribute to cognitive changes.

A neurologist uses the patient's history, neurologic exam, and MRI to determine whether or not a neurodegenerative disease is causing the symptoms at which point the diagnosis of Posterior Cortical Atrophy (PCA) syndrome can be made. However, a syndrome can be caused by many diseases.

Understanding the Underlying Disease

If Alzheimer's disease is causing the syndrome by testing for biomarkers, most commonly with a lumbar puncture to sample the fluid around the spine. If cerebrospinal fluid is negative other considerations are made.

If the patient displays signs of Parkinson's disease or other symptoms like REM behavior disorder, a presumed diagnosis of Lewy Body Disease is made.

If the patient has motor problems on one side of the body a presumed diagnosis of Corticobasal Degeneration may be made.

Cruetzfeldt-Jakob Disease is suspected in rapidly progressive patients, often with characteristic findings on physical exam and MRI, and is confirmed with spinal fluid testing.

LEARN ABOUT RELATED CONDITIONS (HTTPS://WWW.BRAIN.NORTHWESTERN.EDU/DEMENTIA/RELATED-CONDITIONS.HTML)

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Posterior cortical atrophy

AlzheimerSociety

Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative disorder that causes damage and deterioration to the back, or posterior, region of the brain. That area of the brain is responsible for processing what and how we see. PCA may affect a person's vision, their ability to read and write, their ability to navigate, and to reach for objects.

About posterior cortical atrophy

Posterior – the back part of the brain
Cortical – the thinking outer layer of the brain
Atrophy – shrinkage of that area of the brain

PCA is a rare form of dementia that is caused by the abnormal accumulation of amyloid plaques and neurofibrillary tangles in the brain. This build-up of plaques and tangles causes areas of the brain to become increasingly damaged and to shrink (atrophy) over time.

The part of the brain that is most affected by PCA is the posterior cortex, which is the back region (posterior) of the outer layer of the brain (the cortex). As the posterior cortex is responsible for processing visual information, PCA may significantly impair a person's visual functioning.

Since the presence of plaques and tangles in the brain is also a hallmark feature of Alzheimer's disease, PCA is referred to as a variant of Alzheimer's disease.

What are the symptoms?

Symptoms of PCA are most commonly seen in people in their 50s and 60s, but can affect people until their 80s. Early symptoms of this disease are often related to difficulties with a person's ability to process visual information.

Some of the common visual processing symptoms of PCA include: blurred vision, challenges with reading and writing, filling out forms, seeing objects in front of them, and problems with depth perception.

Other symptoms such as difficulties with simple math, knowing how to use tools and appliances, and knowing geographic directions may occur early in the disorder but unlike Alzheimer's disease, short-term memory is not a common symptom in the early stages of PCA. People in the early stages may experience increased anxiety, possibly because they are aware that something is wrong but have difficulties in explaining their symptoms.

Changes in cognitive abilities may be experienced as PCA progresses and causes more damage throughout the brain. This progression will often result in the onset of symptoms that are typical of Alzheimer's disease, like word finding and short-term memory loss.

How is posterior cortical atrophy diagnosed?

A diagnosis may take some time as people will often consult an eye doctor first because they think the symptoms they are experiencing are associated with their eyes. It can be challenging to diagnose PCA early in the progression of the disease. Many people are misdiagnosed because of their young age of onset and the unusual symptoms.

There is no single test to diagnose PCA but rather it is the recognition of the pattern of symptoms. Physicians assess the patterns and types of symptoms to help rule out other conditions that could be causing the symptoms, such as a brain tumor, a stroke or a treatable infection. A thorough assessment may also include testing of someone's thinking abilities, imaging of the brain with either a magnetic resonance imaging (MRI) or computerized tomography (CT), blood tests and specialized vision tests.

What are the causes or the risk factors?

Since the presence of plaques and tangles in the brain is characteristic of both Alzheimer's disease and PCA, research has found that in most cases of PCA, the underlying cause of the disease is Alzheimer's disease or another dementia.

Other neurodegenerative diseases, including: Lewy body dementia, Creutzfeldt-Jakob disease and corticobasal degeneration, also have been shown to cause PCA but this is very rare.

Is there treatment?

Currently, there is no known cure for PCA; however, some medications have been found to be effective in treating its symptoms. Cholinesterase inhibitors that are used to improve alertness and cognition in people with Alzheimer's disease can be used in persons with PCA.

Non-medication therapies can also be helpful including mental and physical exercises. Additional therapeutic approaches support people living with PCA to manage symptoms of their disease. Occupational and physical therapies can help to improve daily functioning. For example, an occupational therapist may provide guidance to people with PCA and their families to ensure that they can safely maneuver around their home, given their visual impairments. Practical visual aids, such as a talking clock, help to support the continued independence of people with PCA.

Support is available:

Visit the Alzheimer Society's website at www.alzheimer.ca or contact your local Alzheimer Society.

Additional resources:

- Alzheimer's Australia: http://www.fightdementia.org.au/common/files/NAT/20130912_NAT_HS_AboutDementiaHelpSheet_22.pdf
- Alzheimer's Society UK: http://www.alzheimers.org.uk/site/scripts/documents_info.php?documentID=135
- UCL Dementia Research Center: http://www.ucl.ac.uk/drc/pcasupport
- UHN Multidisciplinary Memory Clinic: Visual dysfunction in dementia, Home safety recommendations http://www.ucl.ac.uk/drc/pcasupport/contact_links/home_safety_tips

Two closed groups are available on Facebook. These groups are comprised primarily of PCA caregivers, and in some case the actual individual with PCA. Membership is available for people with PCA, their family, friends, colleagues, medical practitioners and support groups.

Contact the Group administrator for access. The groups are:

1) Posterior Cortical Atrophy-early stages
2) Posterior Cortical Atrophy Awareness

The content of this document is provided for information purposes only, and does not represent advice, an endorsement or a recommendation, with respect to any product, service or enterprise, and/or the claims and properties thereof, by the Alzheimer Society of Canada. This information sheet is not intended to replace clinical diagnosis by a health professional.

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R300-38E 2015

What is Posterior Cortical Atrophy?

About the condition

Posterior Cortical Atrophy (PCA – also known as Benson's syndrome) is a progressive degenerative condition which causes damage to brain cells at the back (posterior) of the brain. This area is responsible for processing what we see.

In the vast majority of cases, the loss of brain cells is associated with the same brain changes seen in Alzheimer's disease, namely plaques and tangles. However, it can also be due to other diseases including dementia with Lewy bodies and Creutzfeldt-Jacob disease.

Whilst caused by the same disease process, the effects of PCA and Alzheimer's disease upon the behavior and thought processes of people with each condition are very different. Alzheimer's disease is most commonly associated with deterioration of memory and a progressive decline in language, perception and planning skills. People with PCA tend to have well preserved memory and language but show a decline in vision and experience difficulty with performing skilled movements and with reading and writing. This is due to the brain not being able to interpret and process information received from the eyes.

Changes that might happen

People with PCA generally experience some of these changes associated with vision

- difficulty recognising familiar objects and faces
- increased sensitivity to bright lights or shiny surfaces
- double vision
- particular difficulty seeing in fading or low light conditions
- difficulty judging distance/speed/ perspective – this can cause difficulty with stairs, the speed of traffic, reaching out for things or putting things down on surfaces
- stationary objects may appear to move.

Some changes are hard to understand such as being able to see small print easier than large print, or objects that are right in front of the person not being seen or recognised and then suddenly being seen. This can cause frustration for the person with PCA and their supporters if the changes are not fully understood.

Other changes include:

- difficulty reading, writing, emailing, texting – this can include spelling, and remembering the shape and name of letters or numbers
- difficulty with mental arithmetic and dealing with money and change
- difficulty with coordination and making gestures e.g. waving,
- difficulty using particular tools i.e. cutlery, scissors, glasses
- difficulty with dressing / undressing (particularly zips, buttons, ties, putting arms in sleeves, legs in trousers)
- difficulty finding the chair when sitting down.

Things to consider

Although there is no treatment specific to PCA, people with the condition may find some of the medications used to treat Alzheimer's disease helpful. These medications include donepezil (Aricept), Just like Alzheimer's PCA progresses from one stage to the next and these include:

Stage One: No Impairment (Normal Function)

During this stage, most people will not experience any visual, cognitive, or memory issues.

Stage Two: Very Mild Cognitive Decline

At this stage, an individual may experience subtle problems with various complex visual tasks such as:

- Challenges assembling novel objects/devices
- Loss of confidence in driving abilities owning to the fact that it becomes harder to judge distances
- Issues with writing down or recalling telephone numbers
- Challenges using a computer such as being slower with entering information or typing
- Problems with writing, spelling, reading, and arithmetic

During this early stage, the issues may be intermittent which can make it hard to describe. The arising problems might also not be noticeable to family, friends, or colleagues.

Stage Three: Mild Cognitive Decline

Visual problems become more apparent at this stage for the person who has the condition as well as those close to him or her.

Most individuals will stop driving at this point. Some persons will also get a positive diagnosis of early-stage Alzheimer's disease.

Examples of warning signs that are common in the 3rd stage include:

- Having a hard time seeing things that are right in front of a person
- Getting "lost" on a page when reading
- General clumsiness in persons who were not clumsy before such as putting a glass down sideways, and knocking things over, etc.
- Inability to tell the time from a watch or clock
- Difficulty finding things in familiar places like a handbag or cupboard
- Having a hard time dressing and performing other activities like cutting a piece of toast and other activities like using stairs or coordination.
- Problems with dealing numbers, spelling, and writing
- Paying for items with notes to avoid handling change
- Mild memory issues
- Challenges with picking the right word for a conversation
- Anxiety mainly because a person is more aware of the changes that are happening in their life

Stage Four: Moderate Cognitive Decline

Persons with posterior cortical atrophy at the 4th stage may require help with day to day activities like using appliances, dressing, and cooking amongst others. An individual may have a clear sense of purpose in regards to plans and activities but their attempts to achieve the goals are frustrated by small and frequent visual problems.

Most people during this stage will also have trouble navigating familiar and unfamiliar environments.

Stage Five: Moderately Severe Cognitive Decline

During this stage, the vision continues to deteriorate even further to an extent that a person views the world as different pieces of a puzzle or through the lens of a fractured mirror.

By the end of this stage, some people will be registered as blind implying that they will need support in visually-guided activities.

Stage Six: Severe Cognitive Decline

Considered one of the later stages of PCA, it is normally characterized by impairment of memory, perception, attention, language, and decision-making. Vision impairment remains the most pronounced.

Stage Seven: Severe Cognitive Decline

In the last PCS stage, the problems that affected individual experiences are usually similar to those that affect persons with Alzheimer's. Persons may lose their ability to respond to the environment. They may lose their ability to carry out conversations and eventually affected individuals cannot control movement.

I have received a PCA diagnosis. What next?

A A A

When given a diagnosis of PCA, people naturally ask what will happen next. What challenges might they face and when? What support might be available?

On this page we have provided information on some of the early symptoms commonly associated with PCA as well as available support.

While we try our best to provide general guidance on how abilities may change during the course of PCA, it is important to bear in mind that not everyone will have the same symptoms or progress at the same rate. The experience of PCA will vary according to factors including age, general health and the disease underlying the PCA diagnosis.

Early symptoms

The first symptoms often include difficulties with seeing what and where things are (such as when driving or reading) and with certain everyday tasks.

Vision may be affected in the following ways:

- Problems with reading (for example, difficulty following lines of text, words jumbling up, or with certain text formats and sizes)
- Difficulties with coordination and spatial awareness (such as picking up objects, dressing, selecting the correct numbers on a telephone)
- Visual problems (including perceiving objects amongst clutter, double vision, difficulty focusing)
- Issues with judging distances and speeds (for example with moving traffic, walking onto escalators, descending stairs)
- Issues with recognising faces (including TV characters, friends, relatives)
- Light sensitivity and unusual colour experiences (such as washes of colour when driving at night)

PCA can affect people in different ways and vision is not always the first, or only, area of difficulty for people living with PCA.

Literacy, numeracy and certain coordination skills may also be affected and cause difficulties with the following tasks:

- recalling the exact spelling of words
- handwriting
- remembering the shape or name of particular letters or numbers
- mental arithmetic
- dealing with money and small change
- making gestures (such as waving or giving thumbs up)
- using particular tools, kitchenware or implements (for example scissors, cutlery, glasses)
- getting dressed

As the world mourns the loss of the genius that was Sir Terry Pratchett, a light is being shone on the type of Alzheimer's disease he suffered from.
Sir Terry, who was very vocal about the disease after being diagnosed in 2007, had posterior cortical atrophy (PCA), also known as Benson's syndrome.
The progressive degenerative condition involves the loss and dysfunction of brain cells, and is thought to be behind 5% of cases of Alzheimer's representing thousands of people in the UK.
But although both diseases involve the loss and dysfunction of brain cells, they affect different parts of the brain.

Typically Alzheimer's disease first impacts the sides of the brain - areas which play an important role in memory, which is why memory problems are usually the first symptom of Alzheimer's.
In PCA, the disease first affects the back (posterior) of the brain, known as the occipital lobe.
Professor Sebastian Crutch, an expert on Alzheimer's at University College London (UCL), said this part of the brain was responsible for vision, so people with PCA often initially experienced visual problems.
"Rather than being based on memory where people forget appointments and where they've put things, for people with PCA the first thing they notice is complex visual behaviour," he said.
"They might find they're clipping wing mirrors when they're driving or having problems parallel parking.
"When they're reading, words will jump around and a lot of people will have difficulty perceiving objects such as glass doors."
These are best described as a "brainsight" problem than an "eyesight" issue, he added.

The first symptoms of PCA tend to occur when people are in their late 40s, 50s or early 60s, so it has an earlier onset than Alzheimer's disease.
But the first signs are often subtle and so it may be some time before a formal diagnosis is made.
Prof Crutch said the number of people with PCA was hard to say because it was so difficult to establish.
As damage in the brain spreads and the disease progresses, people develop the typical symptoms of Alzheimer's disease such as memory loss and confusion.
There is no specific treatment for PCA but some people use medications used for Alzheimer's disease.
Sir Terry announced he had the condition in December 2007 when he was 59.
He had been told by doctors that previous summer that he had suffered a mini-stroke at some point in the previous few years, although he had been unaware of it.
After problems with his dexterity and hand-eye co-ordination, he had a series of scans and was diagnosed with PCA.

Dr Clare Walton, research manager at the Alzheimer's Society, said PCA was the "most confusing and poorly understood types of dementia".
"The changes in the brain are very similar to those that happen in Alzheimer's," she said.

"The same changes in brain cells happen, the same proteins build up, and they have the same pathology, but we simply don't know why it affects different parts of the brain."

She said post-mortem examinations show that PCA sufferers' brains "have all the hallmarks of Alzheimer's".

Although it is possible to stabilise people's symptoms, as with Alzheimer's there is no cure.

"Until we have better treatments that stop it from progressing there's not a whole lot we can do," she added.

In a comment piece he wrote on assisted dying for the Guardian in February 2010, Sir Terry described PCA as giving him the "opposite of a superpower".

"Sometimes I cannot see what is there," he wrote.

"I see the teacup with my eyes, but my brain refuses to send me the teacup message. It's very Zen. First, there is no teacup and then, because I know there is a teacup, the teacup will appear the next time I look.

"I have little work-arounds to deal with this sort of thing - people with PCA live in a world of work-arounds."

He described how PCA "manifests itself through sight problems and difficulty with topological tasks, such as buttoning up a shirt".

"If you did not know there was -anything wrong with me, you would not know there is anything wrong with me," he wrote.

"The disease moves slowly, but you know it's there."

Sir Terry likened the condition to a "very, very slow-motion car crash".

"There's an occasional little bang, a crunch, a screw pops out and spins across the dashboard as if we're in Apollo 13. But the radio is still playing, the heater is on and it doesn't seem all that bad, except for the certain knowledge that sooner or later you will definitely be going head-first through the windscreen."

Posterior cortical atrophy

Assoc Prof Frank Gaillard ◉ ◈ et al.

Posterior cortical atrophy, also known as **Benson syndrome**, is an uncommon neurodegenerative disease typically affecting individuals in the sixth and seventh decades of life. It is characterized by dysfunction of the parietal, posterior temporal and occipital lobes, resulting in progressive apraxias, problems with visuospatial and visuoperceptual perception as well as literacy deficiencies [4].

On this page:

Article:

- Terminology
- Epidemiology
- Clinical presentation
- Pathology
- Radiographic features
- Treatment and prognosis
- History and etymology
- Differential diagnosis
- Related articles
- References

Images:

- Cases and figures

Terminology

As is the case with many neurological diseases, the literature is replete with variable terminology. Although generally thought of as a variant of Alzheimer disease (and thus sometimes referred to as a **visual variant of Alzheimer disease**) it is now recognized as sometimes being caused by other pathologies (see below) [4]. As such posterior cortical atrophy should be considered a clinical syndrome with variable etiology.

Epidemiology

Posterior cortical atrophy is generally diagnosed in the sixth and seventh decades (50-65 years of age) with no sex predilection [4].

Incidence is unknown largely in part to the lack of general awareness of the condition and lack of consistent diagnostic criteria. As such the entity is likely under-recognized [4].

Clinical presentation

Posterior cortical atrophy is clinically dominated by disruption of normal higher-order visual proces as such patients eventually behave like individuals who are blind. Patients typically present with [4]:

Privacy - Terms

- visual agnosia: early and pronounced feature
- apraxia: early and pronounced feature
- prosopagnosia
- alexia (difficulty reading)
- environmental disorientation
- Balint syndrome
  - simultanagnosia
  - oculomotor apraxia
  - optic ataxia
- Gerstmann syndrome
  - acalculia
  - agraphia
  - finger agnosia
  - left/right disorientation
- various positive perceptual phenomena e.g. reverse size phenomena, upside-down phenomena etc.

Approximately 25% of patients with posterior cortical atrophy will also develop visual hallucinations [1]. These patients may represent a distinct subgroup, with hallucinations believed to be due to the complex interplay between the midbrain, thalamus and primary visual cortex, rather than the visual association areas [1].

Although mild memory impairment is often present early in the disease, it is clinically different from that seen in Alzheimer disease.

Pathology

In most individuals with posterior cortical atrophy, histology demonstrates focal degeneration with the presence of neuritic plaques and neurofibrillary tangles, especially in parieto-occipital and temporo-occipital areas, and less commonly in the primary visual cortex [4]. As such, posterior cortical atrophy shares features with Alzheimer disease, so much so that some authors believe it to be a variant of the latter, and refer to it as a visual variant of Alzheimer disease. Indeed pathologically the conditions are essentially indistinguishable, other than the distribution of pathological changes and age of onset.

It should be noted that in some individuals who present with posterior cortical atrophy, other underlying etiologies are identified including dementia with Lewy bodies, corticobasal degeneration and even prion disease [4].

Radiographic features

CT SCAN , MRI , SPECT

MRI is the modality of choice for assessing patients with neurodegenerative diseases, although CT may allow gross volume changes to be appreciated. Nuclear medicine functional studies are also of benefit [4].

MRI

The main findings are bilateral, but often more pronounced right-sided, parietal and parieto-occipital and temporo-occipital atrophy. Hippocampi are relatively normal.

Nuclear medicine

SPECT and PET demonstrate hypoperfusion and hypometabolism in the same areas as affected by atrophy and these changes may precede morphological change [4].

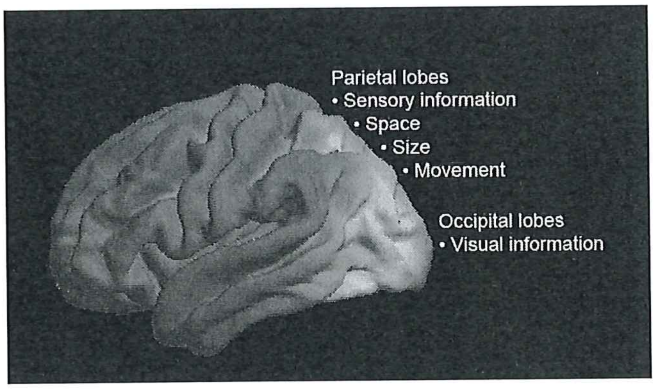
Treatment and prognosis

Privacy - Terms

Before I joined Alzheimer's Society, I carried out research into posterior cortical atrophy - the rare form of Alzheimer's that Terry Pratchett was diagnosed with.

In this post I describe what it is, how it affects people, and what research is taking place. Just to warn you, the post discusses the end stages of Alzheimer's disease which some may find upsetting.

Terry Pratchett had a type of dementia that affects vision, not memory



For many people, the first time they heard of posterior cortical atrophy (PCA for short) will have been when Terry Pratchett announced his diagnosis with the condition.

Posterior cortical atrophy literally means 'back of the brain shrinkage'. The back of the brain is where we interpret visual signals from the eyes and posterior cortical atrophy affects the ability of the brain to process these signals.

Symptoms of PCA can include difficulty reading, recognising objects and judging distances. Problems with spelling and calculation are common, as these skills rely on similar areas at the back of the brain.

Memory and language skills are usually less affected in the early stages of the condition, but as the disease spreads to more parts of

Research

Posterior cortical atrophy

Posterior cortical atrophy (PCA), also known as Benson's syndrome, is a rare degenerative condition in which damage the brain, people may develop more typical symptoms of Alzheimer's disease such as memory loss and confusion.

Vision and perception can be affected by other types of dementia too, but in posterior cortical atrophy these are the first and most severe symptoms.

People tend to develop posterior cortical atrophy at a relatively young age, in their mid-50s or early 60s. We don't have exact numbers but there are about 42,000 people in the UK with a form of dementia that started before the age of 65, and of this group only a small proportion are likely to have posterior cortical atrophy.

occurs at the back (posterior region) of the brain. In the vast majority of people, the cause of PCA is Alzheimer's disease.

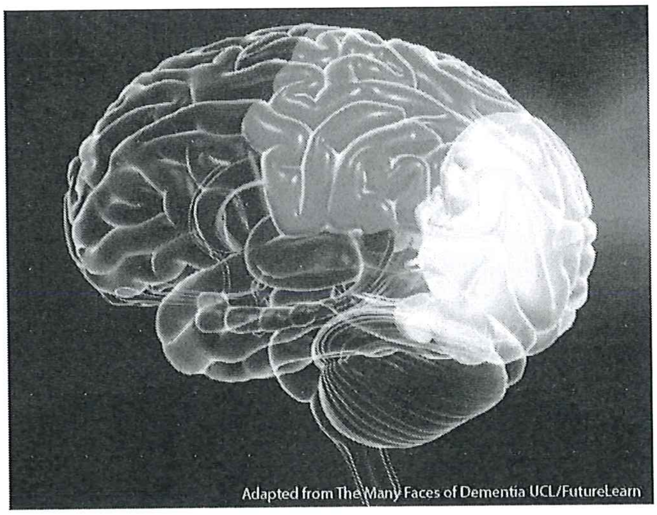
Advice

Young-onset dementia

These pages explain what young-onset dementia is, including the causes and symptoms, and how it is diagnosed and treated.

Raising questions

From a scientific point of view, there's an interesting question that could be answered with research into posterior cortical atrophy. People with posterior cortical atrophy have the hallmarks of Alzheimer's disease in their brain (clumps of toxic proteins called amyloid and tau) but these clumps are found in back of the brain. In Alzheimer's disease these toxic proteins are found in the memory centres, which is why memory loss is one of the main symptoms associated with this condition.



Adapted from The Many Faces of Dementia UCL/FutureLearn

The reasons behind why Alzheimer's disease can affect different parts of the brain in different people are not understood. There must be some genetic, lifestyle and/or environmental factors that cause the visual areas to be vulnerable or the memory centres to be protected in some people. Finding out what these are could give us important insights into Alzheimer's disease.

It's also important to improve our understanding of the symptoms that people with posterior cortical atrophy experience so that we can support them better.

Some of these symptoms might seem quite unusual, for example some individuals have a disordered sense of balance and they might experience dizziness or lean to one side. Alzheimer's Society has given research funding to Professor Sebastian Crutch at UCL to investigate these balance problems. We hope the project will improve understanding, detection and management of these more unusual symptoms.

A progressive condition

As with other forms of dementia, there is no cure for posterior cortical atrophy, and how quickly the symptoms progress depends on the individual person.

Treatments that work for Alzheimer's disease may help relieve symptoms for some people with posterior cortical atrophy, but over time the disease progresses to affect more aspects of thinking and more aspects of everyday life. Terry Pratchett grappled with the issues around the later stages of Alzheimer's and discussed them in the years following his diagnosis.

The period when someone with dementia is approaching the end of their life is difficult and emotional. People with dementia have the right to dignity and a 'good death' and people may wish to discuss these issues in different ways and at different stages. For those who wish to find out more, there is information about end of life care on our website.

Advice

The progression of Alzheimer's disease and other dementias

This page outlines the characteristics of the early, middle and late stages of Alzheimer's disease and briefly looks at how other forms of dementia progress.

Brain Cortex Atrophy

PCA is characterized by problems with visuospatial skills and less prominent memory loss, whereas LPA is characterized by slow speech, word-finding difficulties, and deficits in sentence repetition [28].

From: Diet and Nutrition in Dementia and Cognitive Decline, 2015

Related terms:

Cognitive Defect, Neurofibrillary Tangle, Apraxia, Dementia, Mild Cognitive Impairment, Alzheimer Disease, Frontotemporal Dementia, Primary Progressive Aphasia, Diffuse Lewy Body Disease

Posterior Cortical Atrophy and Dementia

Kannayiram Alagiakrishnan MD, MPH, ... Maher Saqqur MD, MPH, in Diet and Nutrition in Dementia and Cognitive Decline, 2015

Introduction

Posterior cortical atrophy (PCA) is a neurodegenerative condition with an age-of-onset as early as 50 years. PCA is associated with atrophy of the occipital lobe and parieto-occipital lobe. It typically affects the function in the occipital, occipito-parietal, and occipitotemporal regions of the brain. It is considered as a visual variant of Alzheimer's disease (AD) [1,2]. PCA was first described by Benson et al. [3] as a condition with progressive visuospatial dysfunction and relatively preserved memory, insight, and judgment. It is associated with visual dysfunction in the absence of ocular pathology. An estimated 65% of the PCA patients present initially to the ophthalmologist. There is a mean delay of 4 years between symptom onset and diagnosis [4].

Read full chapter

URL: https://www.sciencedirect.com/science/article/pii/B9780124078246000136

Agnosia☆

L.A. Baugh, ... J.J. Marotta, in Reference Module in Neuroscience and Biobehavioral Psychology, 2017

Posterior Cortical Atrophy

Posterior Cortical Atrophy (PCA), also referred to as Benson's disease or the visual variant of Alzheimer's disease, is a progressive neurodegenerative disorder characterized by prominent higher-order visual dysfunction, with preserved memory and reasoning in the early stages of the disorder, but later an insidious, often presenile onset occurs. The initial symptoms of PCA can vary broadly (e.g., achromatopsia, prosopagnosia, object agnosia, environmental agnosia, alexia, agraphia, left-right disorientation, optic ataxia, magnetic misreaching, oculomotor apraxia, dressing apraxia, visual neglect, and simultanagnosia). However, early onset symptoms are often related to difficulties with a person's ability to process visual information. For example, PCA patients often demonstrate impairments in global processing or the ability to view the visual world as a coherent whole. Traditionally, this deficit in PCA has been labeled as simultanagnosia. The responses of individuals with PCA to basic perceptual tasks, such as object identification using line drawings, suggest that they are unable to take into account all of the available information when viewing an image. PCA patients persistently attempt to identify images based on very select information extracted from a small part of the overall picture. When viewing faces, for example, there is often a focus on nondiagnostically useful locations suggesting that individuals with PCA have difficulty processing the face at either the featural or configural level. In fact, it appears that the decreased effective field of view in PCA patients is so severe that it results in an extreme dependence on local processing, such that a feature-based approach is not even possible.

PCA is characterized by progressive bilateral atrophy in the posterior areas of the brain (e.g., occipito-parietal and occipito-temporal areas), often with a predominance in the right hemisphere. In the majority of cases, Alzheimer's pathology is the underlying cause, but cases have been documented as a result of corticobasal degeneration, dementia with Lewy bodies, and prion diseases such as Creutzfeldt-Jakob disease.

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URL: https://www.sciencedirect.com/science/article/pii/B9780128093245002534

Alzheimer's Disease and Dementia

Patrick A. Lewis, Jennifer E. Spillane, in The Molecular and Clinical Pathology of Neurodegenerative Disease, 2019

2.2.3 Posterior Cortical Atrophy

Posterior cortical atrophy is a rare dementia syndrome characterized predominantly by a progressive decline in visual processing skills. The early symptoms may be nonspecific complaints of visual blurring, but symptoms progress to difficulty reading, difficulties perceiving textures and patterns, and an inability to piece together visual scenes to create a global picture that makes sense (simultagnosia) [31]. Most cases of posterior cortical atrophy are related to Alzheimer's pathology, but other pathologies have described too, including Lewy body dementia, corticobasal degeneration, and prion disease [32,33].

Read full chapter

URL: https://www.sciencedirect.com/science/article/pii/B9780128110690000021

Social Cognitive Neuroscience, Cognitive Neuroscience, Clinical Brain Mapping

K.A. Josephs, in Brain Mapping, 2015

Posterior Cortical Atrophy

The term PCA is synonymous with Benson's syndrome (Benson et al., 1988). Patients with PCA present with visual spatial and perceptual difficulties. Commonly patients with PCA have had multiple previous visits to the ophthalmologist due to complaints of vision yet no ophthalmologic abnormalities can be identified. At the bedside, PCA patients show evidence of simultagnosia, optic ataxia, and ocular motor apraxia, consistent with the Bálint's syndrome. Similar to patients with LPA, PCA patients may also have difficulties with calculations. Patients with PCA show long disease duration, sometimes approaching two decades. Over time, patients with PCA will also have difficulty with praxis and language and may therefore have overlapping features with PLX and LPA. Loss of episodic memory in PCA is common over time and may even be mild early on in the disease course. Neuropsychological tests reveal poor performance on visuospatial and perceptual tasks.

Patients with PCA typically show a focal pattern of biparietal atrophy (Whitwell et al., 2007) and hypometabolism (**Figure 1**). In most instances, the right hemisphere is more affected than the left. When the left hemisphere is more affected than the right, language abnormalities may be more obvious. In some instances, parietal involvement may extend into the occipital regions. Overtime, atrophy and hypometabolism become more extensive and may involve the medial temporal and frontal lobes.

Patients with PCA typically do not show any familial inheritance, and hence, no specific gene abnormalities have been associated with PCA. In PCA, over 90% of patients have underlying Alzheimer's type pathology (Renner et al., 2004). Like in PLX and LPA, there tends to be with relative sparing of neurofibrillary tangles in the hippocampus.

Read full chapter

URL: https://www.sciencedirect.com/science/article/pii/B9780123970251000804

The Primate Nervous System, Part II

J.H. Morrison, ... G.W. Huntley, in Handbook of Chemical Neuroanatomy, 1998

9.2.2 Neuropathologic analysis of posterior cortical atrophy cases

Quantitatively, the densities and distribution of senile plaques and neurofibrillary tangles in the posterior cortical atrophy cases that we had the opportunity to study (Hof et al. 1989, 1990b, 1993, 1997a; Hof and Bouras 1991), generally agrees with the finding of other investigators who also reported inordinately high densities of AD lesions in the occipital regions (Berthier et al. 1991; Jacquet et al. 1990; Mendez et al. 1990a,b,c; Graff-Radford et al. 1993; Levine et al. 1993; Victoroff et al. 1994). These observations are summarized in Figures 59 and 60. There is a gradient in neurofibrillary tangle densities from area V1 to the visual association regions in the parietal cortex, as well as in the posterior cingulate cortex in these cases who all suffered from visuomotor impairment resembling Bálint's syndrome, except in one who presented clinically with prosopagnosia and apperceptive visual agnosia (Hof and Bouras 1991). A comparable occipital shift in the lesion distribution is observed in this case, however, it differs from the others in that there is a preferential distribution of neurofibrillary tangles and senile plaques in the inferior temporal cortex (areas TEO and TE), whereas the inferior parietal and posterior cingulate cortex are less affected, demonstrating the specific involvement of certain components of the occipitotemporal visual pathway, consistent with the clinical symptomatology (Hof and Bouras 1991).

Advertisement

Neurology®

August 22, 2019 e-Pearl of the Week: Benson syndrome

Benson syndrome

Benson syndrome or posterior cortical atrophy (PCA), also known as a visual variant of Alzheimer disease presents with visuospatial and perceptual difficulties. Memory, insight, and judgment are preserved in early stages[1]. The exact etiology is unknown with no genetic causation found. The MRI shows atrophy of parietal and occipitotemporal cortices[3]. Management is using voice recognition software, audio books and lighting tools at home. Currently, no data supports the use of acetylcholinesterase inhibitors[2,3]. Patients usually succumb within 8–12 years from the time of onset.

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CONTINUE FIND OUT MORE

Though PCA and Alzheimer's disease both involve the loss and dysfunction of neurons, or brain cells, they affect different parts of the brain.

For the most part, Alzheimer's usually first impacts the sides of the brain, those areas that play an important role in memory, which causes the first symptoms of memory loss.

PCA, first described in 1988 by Dr. D. Franck Benson, develops slowly and first affects the back of the brain known as the occipital lobe, which is responsible for vision. Often, those first experiencing this condition describe visual problems or complex visual behaviors.

Those with PCA symptoms would have difficulty perceiving objects like glass doors, difficulty in recognizing people and can struggle with reading, writing and numerals. Additionally, they might have difficulty with some tasks, such as buttoning up a shirt.

The syndrome is sometimes caused by other diseases, such as dementia with Lewy Bodies.

Signs of depression and anxiety tend to be prominent in the early stages of PCA. Early detection and treatment of psychiatric symptoms can affect function and independence in daily life.

The first symptoms of PCA typically occur in those in their late 40s, 50s or early 60s, and are often subtle and difficult to diagnose. As the brain deteriorates and the disease progresses, more Alzheimer's-like symptoms occur, such as memory loss and confusion.

Risk factors for developing the disease are unknown. PCA is said to be the most poorly understood type of dementia, and there is no specific treatment. Most often the same medications prescribed for Alzheimer's disease are used.

While researchers have observed that PCA and Alzheimer's disease have the same changes in the brain and the same protein buildup and the same pathology, it is a mystery why this specific dementia affects different parts of the brain.

Because of the nature of the disease, PCA presents unique challenges for clinicians who counsel those affected and their families on clinical status and prognosis, and experts designing clinical trial of interventions.

QUESTIONS ABOUT ALZHEIMER'S DISEASE OR RELATED DISORDERS CAN BE SENT TO DANA TERRITO, THE MEMORY WHISPERER, OWNER OF DANA TERRITO CONSULTING, LLC, AT THEMEMORYWHISPERER@GMAIL.COM.

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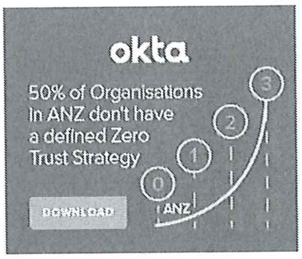
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We Breakdown What Never Trust Adoption Looks Like:...
By **Okta**
Okta dug in and surveyed security leaders about their current Zero Trust projects.

beginning between the ages of 50 years and 65 years.[3] The underlying pathology in the majority of individuals is Alzheimer's disease (AD), although a small number of cases attributable to Lewy body disease and corticobasal degeneration have been reported.[4 5] The prevalence is unknown. PCA is characterised by initial neurodegeneration towards the back of the brain, specifically in the parietal, occipital and occipitotemporal regions.[6] Correspondingly, the initial symptoms predominantly relate to cortical visual impairment, particularly deficits in visuospatial and visuoperceptual processing. Other characteristic symptoms relate to impairments associated with posterior functions, including literacy, spelling and numeracy.[6 7] While current clinical criteria cite visual processing impairments with proportionally less impaired memory as core diagnostic features of PCA, patients may in fact exhibit memory impairments at initial presentation.[4 8 9] When compared with other dementias, PCA is relatively under-researched. The majority of research into PCA is concentrated around establishing the neuropsychological, cognitive and imaging profile(s) of those with the diagnosis.[4 9–11] Anecdotal and laboratory-based evidence suggests ways in which PCA might impact on people's daily lives, including problems with reading, driving and localising objects in space,[9 12–14] but there is a paucity of research focusing on the everyday impact of living with the diagnosis for individuals and families.

One recent paper investigated the impact of PCA on activities of daily living, documenting difficulties with performance of everyday skills for people with PCA (including operation of appliances, writing and handling money) and self-care (including dressing, feeding and bathing) compared with predominant deficits in memory, motivation and orientation of people with typical, amnestic presentations of AD (tAD).[15] In another recent paper, Suárez-González *et al*[16] investigated the neuropsychiatric profile of people with PCA. They found similar increases in depression, apathy, irritability and anxiety for those with PCA to participants with tAD.

In the absence of disease-modifying therapies for AD and other forms of dementia, environmental and psychosocial interventions to improve the quality of life and well-being of those living with it hold particular significance.[17–19] Better understanding the needs of those with different rare and/or young-onset dementias will be an important step in developing effective environmental or psychosocial interventions. A second generation of literature is beginning to delineate the particular experiences of those with less common forms of dementias such as young-onset dementias[20 21] and those with atypical symptom profiles like behavioural variant frontotemporal dementia.[22] Developing a greater understanding of the day-to-day impact of dementia in relation to visual problems will be a timely addition to this, not least because those with tAD may also go on to have cortical visual impairment, likely later on in their diagnosis and as such at a time when they may not be so able to articulate their experiences of the symptoms.[23]

These varied everyday impacts of dementias that pose challenges for quality of life and well-being are often considered within a broad conceptual category of stressors, defined as demands that are considered to exceed a person's available resources. Cognitive, emotional and/or behavioural attempts to manage these demands are often approached and studied as coping strategies. Much attention has consistently been given over the past few decades to understanding what contributes to informal caregiver stress and what facilitates coping, given the huge societal contribution informal carers make by continuing to care for loved ones with dementia at home.[24 25] The stress-coping approach is widely acknowledged as dominant within this literature,[26 27] and its popularity is exemplified by the numerous reviews into stress and coping in dementia noted by Gilhooly *et al*.[19] There are multitudes of studies looking at the chronic and particular psychological, socioemotional and practical stressors and strains faced by carers of people with dementia[28 29] and also the nature and efficacy of the many practical, emotional, psychological and social coping strategies employed to mediate this.[30 31]

The current study sought to maximise on the relative abilities of those with PCA to reflect on and communicate their experiences. Using the Stress Process Model[32 33] as a conceptual framework, we present findings from a qualitative exploration of the stresses associated with mild to moderate stage PCA and responses to these over time. The Stress Process Model outlines primary stressors that result directly from the disease itself, secondary strains that may follow and both internal and external factors that mediate both of these in shaping outcomes. Having been developed to conceptualise informal caregivers' experiences,[32] it has since been adapted for individuals with dementia,[33] and in doing so acknowledges the multiple perspectives to be taken into account when understanding the dementia experience. More specifically, the study aimed to explore the potential of the physical environment to contribute to and/or mediate the stress process owing to the prominent visuospatial and visuoperceptual symptoms.

METHODS

Design/sampling

A qualitative design was deemed appropriate in order to gain rich detailed accounts and for possibly unanticipated insights to emerge.[34] This was considered important because of both the paucity of knowledge about the impact of PCA on individuals' subjective experiences and also the distinctiveness of the symptom profile. In-depth semi-structured interviews were chosen owing to the abilities of those with PCA to recount their experiences and, again, to facilitate the collection of data of sufficient richness and depth. Participants were recruited via the Specialist Cognitive Disorders Clinic at the National Hospital for Neurology and Neurosurgery, University College London Hospitals NHS Foundation Trust. Inclusion criteria were a

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Posterior cortical atrophy

Posterior cortical atrophy (PCA), also known as Benson's syndrome, is a rare degenerative condition in which damage occurs at the back (posterior region) of the brain. In the vast majority of people, the cause of PCA is Alzheimer's disease.

Posterior Cortical Atrophy: Do I see what you see?

Do you know what Posterior Cortical Atrophy is? Watch Lori's...

19 April 2018 🗨 8

The first symptoms of PCA tend to occur when people are in their mid-50s or early 60s.

However, the first signs are often subtle and so it may be some time before a formal diagnosis is made.

Initially, people with PCA tend to have a relatively well-preserved memory but experience problems with their vision, such as difficulty recognising faces and objects in pictures. They may also have problems with literacy and numeracy.

These tasks are controlled by the back part of the brain, where the initial damage in PCA occurs.

As damage in the brain spreads and the disease progresses, people develop the more typical symptoms of Alzheimer's disease, such as memory loss and confusion.

There are no specific medications for the treatment of PCA but some people find medications for Alzheimer's disease helpful.

Posterior Cortical Atrophy

What is Posterior Cortical Atrophy?

Posterior Cortical Atrophy, or PCA, is most commonly an atypical clinical presentation of Alzheimer's disease. People with PCA often come to the clinic because they are having trouble interpreting information they see with their eyes (known as "visual processing impairment"), rather than with symptoms of memory loss. Because memory loss is the most common presenting symptom of Alzheimer's disease, PCA is referred to as "atypical Alzheimer's disease."

What is the visual variant of Alzheimer's disease? Is it the same as PCA?

The visual variant of Alzheimer's disease, or VVAD, is a term that can be appropriately applied to PCA in certain circumstances, such as when a person experiences dementia due to Alzheimer's disease and the predominant symptom is visual processing impairment. However, the diagnostic term of VVAD is falling out of favor because we have new criteria to support a diagnosis of PCA that would include VVAD.

Is PCA always due to Alzheimer's disease?

No. It is estimated that PCA is due to causes other than Alzheimer's disease approximately 15% of the time. Other causes of PCA include Lewy body dementia and corticobasal degeneration.

How and why is vision affected in PCA?

The eyes themselves are not affected by PCA, yet people with PCA have a difficult time seeing and interpreting visual information. Symptoms vary with each person, but the most common presentation at onset is simply having problems seeing without an explanation. As the disease progresses, symptoms become more apparent and often include difficulty with depth perception, problems finding objects that are in plain view, difficulty reading, trouble driving, and problems recognizing faces. It should be noted that the formation of some new memories relies on our ability to process visual information. Therefore, PCA can contribute to problems remembering visual information because the brain did not properly process it.

The reason that vision is affected in people with PCA is because at least 25% of the human brain has vision and visual processing as its main responsibility. The regions in the back of the brain (known as the posterior regions) are where most of the visual processing takes place, thus the word "posterior" in PCA. The part of the brain affected is the "cortex," which is the layered and wrinkled part of the brain that processes information at a very high level. Since the brain cells in the posterior cortex are lost to the disease, the term "atrophy" is used, which refers to the loss or shrinking that occurs when brain cells die. Although PCA was named because of the distinctive atrophy of the cortical regions in the back of the brain observed by an MRI scan, atrophy of the posterior regions of the cortex may not be prominent when symptoms first appear.

How is PCA diagnosed?

There is no one test that can be used to make the diagnosis of PCA. Instead, complete eye and neurologic examinations are necessary to make sure there are no other causes of the visual symptoms, and basic laboratory tests and brain imaging are also important components of the evaluation. Often, visual field tests, which measure the visual sensitivity of the full field of vision, can show problems not explained by an MRI of the brain or by the examination of the eye itself and can lead to the discovery of PCA. Visual field tests are often ordered by ophthalmologists or optometrists when trying to determine the cause of a person's visual problem when an examination of the eye does not allow for a diagnosis. Also important to the diagnosis are the cognitive tests of attention, memory, language, and executive functions that are helpful in differentiating PCA from other problems.

Is there a treatment for PCA?

There is currently no treatment for PCA, but supportive measures and symptomatic medications, such as those used in the treatment of Alzheimer's disease, and other therapies may help. The CU Alzheimer's and Cognition Center at the University of Colorado School of Medicine has on-line information about Alzheimer's disease. Dr. Victoria Pelak directs patient support group meetings every other month for people with PCA, and you can find out more about this by clicking here: Colorado PCA Support. Another important on-line site, with video-taped lectures about PCA and information and support can be found at http://pca-vision.org/ If you have further questions, please see our Colorado PCA Support webpages where you will find a link to email us with your questions.

Additional Navigation ⌄

HOMEPAGE / POSTERIOR CORTICAL ATROPHY (PCA) / UNDERSTANDING PCA

Understanding PCA

Posterior cortical atrophy (PCA) is a form of dementia that mainly affects the parts of the brain that process visual and spatial information.

A A A

Common first signs and symptoms include difficulties with seeing what and where things are (for example, when driving or reading). This includes:

- problems with reading
- issues with recognition
- difficulty with coordination
- issues with judging distances
- light sensitivity

The symptoms of PCA usually begin before the age of 65 and and people are often in their mid-50s or early 60s when they first experience symptoms. However, PCA can also affect older people and it can take a long time for people to receive the correct diagnosis.

Causes

Posterior cortical atrophy (PCA) means 'back of the brain shrinkage' and it refers to the progressive loss of brain cells, particularly in brain regions that process visual and sensory information, such as the occipital and parietal lobes. The changes in the brain that cause PCA can be caused by different disease processes. In the majority of cases PCA is caused by changes to brain cells similar to those that occur in Alzheimer's disease.

However, the initial symptoms of PCA are very different from those of Alzheimer's disease. This is because of the different areas of the brain that are affected. In Alzheimer's disease, the 'memory areas' of the brain are particularly affected, whereas in the early stages of PCA the skills controlled by the rear of the brain (such as vision) are more affected.

People living with PCA often preserve their memory and language abilities in the early stages, but experience a progressive decline in vision and/or literacy skills (including spelling, writing and arithmetic).

There are a small number of underlying conditions, as well as Alzheimer's disease, that can cause PCA:

These include:

Lewy body dementia (LBD) – people may experience visual hallucinations, and may become stiffer and slower in their movements, in a similar way to people living with Parkinson's disease.

Corticobasal Syndrome (CBS) – people may have difficulty using one side of their body or experience jerky, awkward or slow movements.

Prion diseases (e.g. Creutzfeldt-Jakob disease (CJD)) – very rare conditions in which people may experience a very rapid decline in their cognition.

Very little is known about why PCA affects some people and not others, nor how many people there are living with PCA. Research is leading to improved understanding and you can learn more about ongoing **PCA research** here.

Diagnosis

The first signs of PCA are often very subtle and may be difficult for the person experiencing them to describe.

People with PCA usually first experience difficulties with vision. Quite understandably, this leads them to consult an optician. However, the visual problems in PCA are due to changes in the brain rather than the eyes, and so an optician will not usually get to the root of the problem. In PCA the problem is that the affected part of the brain is unable to interpret the information sent to it by the healthy eyes.

Problems with spelling, numeracy and learned motor skills have also been reported as early indicators of PCA.

Even once an appropriate referral has been made to a specialist – usually a neurologist, psychiatrist or neuro-ophthalmologist – it may take some time before the diagnosis is made formally. It can typically take 1 to 3 years from the onset of symptoms. As a result, many people living with PCA feel frustrated by the time of diagnosis.

It is important that opticians, ophthalmologists, GPs and others across the healthcare sector are better informed about PCA and better equipped to recognise the symptoms. Rare Dementia Support is working to improve understanding of PCA and other rare dementias and there are some useful resources available on our **Learning page**. We have created a short animated film, narrated by people living with PCA and their partners. Watch film '**Do I see what you see?**'.

By increasing awareness and understanding of PCA, we hope to ensure that healthcare professionals and communities are better equipped to support people affected by a diagnosis of PCA. Help us by sharing your stories and contacting us at **contact@raredementiasupport.org**

Testing

There is no single diagnostic test for PCA. The tests listed below may help to exclude potentially treatable causes such as infection, inflammation or brain tumours:

- specialised visual tests (organised by eye-specialists)
- neuropsychological tests of cognitive skills (including memory, visual perception, literacy)
- blood tests
- brain scans may reveal shrinkage of the back part of the brain as a result of brain cell loss (for example, magnetic resonance imagery (MRI) or computerised tomography (CT))
- lumbar puncture (examination of the fluid circulating around the brain and spinal cord)
- EEG (recording of the electrical activities of the brain)

More information can be found in our **FAQs section**.

Living with PCA: A short film

What happens as PCA progresses? (sensitive)

Professor Sebastian Crutch describes how posterior cortical atrophy changes over time, and the seven stages of PCA project.

The Stages of PCA project is based on a framework developed for typical AD, and aims to give an idea of the different symptoms that may be experienced at different stages. There are many caveats to the stages, everybody is different and people may move at a faster or slower rate; and people's perception of what is 'mild' and what is 'severe' may be different.

We've listed the labels of the stages with a couple of aspects for each stage below, but do take a look at the stages of PCA document (see downloads below) to see all the caveats and more detail about each stage.

Stage 1. No impairment (normal function)

Stage 2. Very mild cognitive decline
- Difficulties with reading and writing
- Difficulty using a computer

Stage 3. Mild cognitive decline
- Difficulty finding things in a handbag, cupboard, etc.
- Difficulty seeing things which are 'right under your nose' or only seeing part of something

Stage 4. Moderate cognitive decline
- Inability to read labels on food packets
- Problems discriminating coins

Stage 5. Moderately severe cognitive decline
- Require help with most or all everyday tasks
- Require support or supervision when walking due to visual difficulties or movement difficulties

Stage 6. Severe cognitive decline
- May become 'functionally blind' requiring support in all visual activities
- Experience deterioration of other sensory functions such as touch

Stage 7. Very severe cognitive decline
- Symptoms may resemble typical AD more closely than at previous stages
- Individuals may have a reduced ability to respond to their environment and carry on a conversation

In the video we heard Professor Crutch explain how different people like to receive different information about the disease at different times. What are the things that would encourage you to want more information earlier, and what are the things that would mean you prefer not to find everything out right away?

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MeSH: -

GARD: -

MedDRA: -

Summary

Epidemiology

Posterior Cortical Atrophy (PCA) prevalence is unknown, largely due to the lack of awareness of the syndrome, delayed diagnosis and the variable terminology referring to it (now partially addressed through international consensus criteria).

Clinical description

Early PCA symptoms include visuoperceptual and visuospatial dysfunction, apraxia and alexia. Features of Bálint syndrome (simultanagnosia, optic ataxia and oculomotor apraxia) and Gerstmann syndrome (acalculia, agraphia, finger agnosia and left-right disorientation) are commonly described. The disorder typical onset is between 50-65 years of age. Earliest reported symptoms include difficulties with complex visual behaviors (e.g. driving, reading, and telling the time from an analogue watch). Reading problems include getting lost on the page (visual disorientation), overlapping or miscombined letters (visual crowding) or better reading of small than large print. Odd visual manifestations like abnormally prolonged color after-images and perception of movement of static stimuli are also reported. People with PCA tend to have relatively well preserved memory, insight, and judgment early in the disease course, though language (word finding problems, phonological errors) often emerge early. It should also be noted that PCA is a relatively heterogenous syndrome with some experiencing a very focal visual syndrome, whilst others show a more mixed cognitive picture with prominent visual but also evident episodic memory deficits from early in the disease course. Individuals with PCA often experience anxiety and depression from early on. Extrapyramidal signs, myoclonus, and grasp reflex have also been reported in PCA patients. An international consensus classification frameworkdistinguishes two syndromic description levels. Classification 1 (PCA) defines the core clinical, cognitive, and neuroimaging features and exclusion criteria of the clinico-radiological syndrome. Classification level 2 (PCA-pure, PCA-plus) establishes whether, in addition to the core PCA syndrome, the core features of any other neurodegenerative syndromes are present.

Etiology

A third level of the international classification framework provides disease-level descriptions of the underlying causes of the PCA syndrome, based on available pathophysiological biomarker evidence. This distinguishes between Alzheimer's disease as the most common underlying pathology (PCA-AD), and other causes including Lewy Body disease (PCA-LBD), corticobasal degeneration (PCA-CBD) and prion disease (PCA-prion).

Diagnostic methods

PCA is an under-recognized disorder resulting often in a significant delay in diagnosis. Diagnosis of this clinico-radiological syndrome is based on neurological assessment, specific visual and cognitive testing, and (optional supportive) brain imaging and routine blood tests.

MRI characteristically shows bilateral atrophy in the occipital, parietal and posterior temporal lobes, often asymmetric being more pronounced in the right hemisphere. Single photon emission computed tomography (SPECT) or PET show hypometabolism of the posterior cerebral areas as well as in the frontal eye fields in more advanced stages. Conclusive diagnosis of the underlying disease is confirmed on brain autopsy, though CSF and amyloid imaging may help to refine the clinical diagnosis.

Differential diagnosis

Differential diagnosis includes the most commonly associated neuropathology Alzheimer disease, but may also include Lewy body disease, cortico-basal degeneration, and prion diseases such as Creutzfeldt-Jakob disease.

Management and treatment

There is currently no cure for any of the neurodegenerative conditions that most commonly underly the PCA syndrome. Medications like acetylcholinesterase inhibitors, used in the treatment of Alzheimer's disease, are available and might bring relief for some symptoms. Management of PCA is based on visual aids, rehabilitation programs that include psychoeducation, compensatory strategies, and cognitive exercises to cope with visual disabilities. Antidepressant treatments may benefit individuals with PCA who have relatively preserve insight into their decline and often deal with depression, irritability, frustration and a loss of self-confidence. People with PCA and their caregivers are likely to have different needs to people with more typical presentations of Alzheimer's disease, and may benefit from specialized support groups.

Prognosis

Prognosis is poor as PCA is progressive disorder. Life expectancy after PCA diagnosis is thought to be similar (8-12 years) to individuals affected with Alzheimer's disease.

Expert reviewer(s): ***Pr Sebastian CRUTCH*** *- Last update:* ***July 2020***

Detailed information

Professionals

> Review article

English (2012)

Additional information

Further information on this disease

Support (https://www.alzheimersresearchuk.org/dementia-information/types-of-dementia/posterior-cortical-atrophy/support/)

Risk factors (https://www.alzheimersresearchuk.org/dementia-information/types-of-dementia/posterior-cortical-atrophy/risk-factors/)

Research (https://www.alzheimersresearchuk.org/dementia-information/types-of-dementia/posterior-cortical-atrophy/research/)

What is Posterior cortical atrophy?

PCA is caused by damage to the brain cells at the back of the brain that make sense of what our eyes are seeing. It is also known as Benson's syndrome.

Alzheimer's disease is most often the cause of the brain cell damage in PCA, but it is sometimes caused by other diseases, such as dementia with Lewy bodies. PCA is sometimes called a visual form of Alzheimer's. However, the early signs of PCA and typical Alzheimer's can be very different. Alzheimer's disease usually affects memory first, but in PCA the first signs are often problems with vision and perception.

People often develop PCA at an earlier age than typical Alzheimer's disease, usually between the ages of 50 and 65. PCA is a less common form of dementia, and at the moment we can't be sure how many people around the world are affected by it.

What is posterior cortical atrophy?

Find out more about the symptoms and causes of posterior cortical atrophy, and the treatments currently available.

Download booklet **(/wp-content/plugins/mof_bl_0.2.9/downloads/PCA-0419-0421_WEB.pdf)**

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☎ (tel:03001115555)

✉ (mailto:enquiries@alzheimersresearchuk.org)

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Posterior cortical atrophy

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Posterior cortical atrophy (PCA) is a rare form of dementia that usually begins by affecting a person's vision. It is also known as Benson's syndrome.

About (https://www.alzheimersresearchuk.org/dementia-information/types-of-dementia/posterior-cortical-atrophy/)	**Symptoms (https://www.alzheimersresearchuk.org/dementia-information/types-of-dementia/posterior-cortical-atrophy/symptoms/)**

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Posterior Cortical Atrophy

Listen (https://app.readspeaker.com/cgi-bin/rsent?customerid=8204&lang=en_uk&readid=content&url=https%3A%2F%2Fwww.youngdementiauk.org%2Fposterior-cortical-atrophy)

Posterior Cortical Atrophy (PCA) is the degeneration of the cells at the back or posterior of the brain.

The changes in the brain resemble those in someone with Alzheimer's disease but the effects are very different. People with PCA will often first experience problems with their sight. This is because their brain is having trouble interpreting the information sent to it by the eyes. People may also struggle with spelling, writing or maths.

You will find more detailed information about the condition on these websites R (http://www.ucl.ac.uk/drc/pcasupport)are Dementia Support (http://www.raredementiasupport.org/posterior-cortical-atrophy-pca/) I Posterior Cortical Atrophy Awareness (https://www.facebook.com/groups/147542335356010/?fref=ts) (Facebook group) | Do you see what I see? (https://youtu.be/jekW8Z93LMw) A short film about PCA

Read Valerie's story about living with PCA here (https://www.youngdementiauk.org/valeries-story) and Tracey's story here (https://www.youngdementiauk.org/traceys-story).
In Sarah's story (https://www.youngdementiauk.org/posterior-cortical-atrophy) she talks about life after husband Andy's PCA diagnosis.

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Posterior Cortical Atrophy

Posterior cortical atrophy (PCA) refers to gradual and progressive degeneration of the outer layer of the brain (the cortex) in the part of the brain located in the back of the head (posterior).

About posterior cortical atrophy

It is not known whether posterior cortical atrophy is a unique disease or a possible variant form of Alzheimer's disease. In many people with posterior cortical atrophy, the affected part of the brain shows amyloid plaques and neurofibrillary tangles, similar to the changes that occur in Alzheimer's disease but in a different part of the brain. In other people with posterior cortical atrophy, however, the brain changes resemble other diseases such as Lewy body dementia or a form of Creutzfeldt-Jakob disease. Most cases of Alzheimer's disease occur in people age 65 or older, whereas the onset of posterior cortical atrophy commonly occurs between ages 50 and 65.

Prevalence

There is no standard definition of posterior cortical atrophy and no established diagnostic criteria, and so it is not possible to know how many people have the condition. Some studies have found that about 5 percent of people diagnosed with Alzheimer's disease have posterior cortical atrophy. However, because posterior cortical atrophy often goes unrecognized, the true percentage may be as high as 15 percent. Researchers and physicians are working to establish a standard definition and diagnostic criteria for posterior cortical atrophy (PDF).

Symptoms

The symptoms of posterior cortical atrophy can vary from one person to the next and can change as the condition progresses. The most common symptoms are consistent with damage to the posterior cortex of the brain, an area responsible for processing visual information. Consistent with this neurological damage are slowly developing difficulties with visual tasks such as reading a line of text, judging distances, distinguishing between moving objects and stationary objects, inability to perceive more than one object at a time, disorientation, and difficulty maneuvering, identifying, and using tools or common objects. Some patients experience hallucinations. Other symptoms can include difficulty performing mathematical calculations or spelling, and many people with posterior cortical atrophy experience anxiety, possibly because they know something is wrong. In the early stages of posterior cortical atrophy, most people do not have markedly reduced memory, but memory can be affected in later stages.

Diagnosis

Misdiagnosis of posterior cortical atrophy is common, owing to its relative rarity and unusual and variable presentation. Additionally, people with posterior cortical atrophy frequently first seek the opinion of an ophthalmologist who may indicate a normal eye examination by their usual tests. Because the first problems are perceived as eye problems, cortical brain dysfunction initially may not be considered as a cause.

There are no standard diagnostic criteria for posterior cortical atrophy, although diagnostic criteria are being developed (PDF). Physicians rely on a combination of neuropsychological tests, blood tests, brain scans and a neurological examination to diagnose the condition and rule out other potential explanations for symptoms. Characteristic features that are sometimes used for diagnosis include gradual onset of visual symptoms (described above) with preservation of normal eye function and preservation of memory. Age of onset between 50 and 65 years is another clue suggesting PCA. The diagnosis should rule out the possibility that the symptoms were caused by a stroke, tumor or other identifiable condition.

There is an ongoing discussion in the field whether posterior cortical atrophy should be considered a form of Alzheimer's disease or a distinct disease entity. Brain imaging has shown that the posterior cortex is thinner in people with posterior cortical atrophy than healthy people of the same age. This indicates that the individual has experienced a decrease in brain volume. Furthermore, people with posterior cortical atrophy have degeneration in different parts of the brain than people with typical forms of Alzheimer's disease, although there is often overlap between the two conditions.

Causes and risks

Similar to Alzheimer's disease, the causes of posterior cortical atrophy are unknown, and no obvious genetic mutations have been shown to be linked to the condition. It is also not known if the risk factors for Alzheimer's disease are also risk factors for posterior cortical atrophy.

Treatment and outcomes

There are no treatments for posterior cortical atrophy known to slow or halt its progression. Because posterior cortical atrophy resembles Alzheimer's disease in some patients, it has been suggested that drugs used to temporarily alleviate brain dysfunction in Alzheimer's disease may be helpful in posterior cortical atrophy, but this is not proven. Some people with posterior cortical atrophy may benefit from treatment to alleviate symptoms such as depression or anxiety, but the overall benefits and risks of such treatments are not established.

Alzheimer's Association Research

Traumatic Brain Injury and Chronic Traumatic Encephalopathy (CTE) Awardees

Year	Researcher	Study Name
2015	Jesse Mez	Exploring MAPT and Tau-Related Genes in Chronic Traumatic Encephalopathy
2014	Thor Stein	Genetic Risk Factors Underlying Chronic Trauma and Alzheimer's Disease Patholog
2014	Kun Ping Lu	Validation of cis-Tau as a Therapeutic Target for Alzheimer's Disease
2014	Tsuneya Ikezu	Exosome Pathway as a Novel Therapeutic Target of Tauopathy
2013	Eva De Lago	Role of the Endocannabinoid System in TDP43-related Dementia
2013	Andrew Watt	Characterisation of Tau Imaging Ligands for Alzheimer's Disease and other Dement
2010	Marco Prado	The Prion Protein as a Therapeutic Target in Alzheimer's Disease
2007	Michael Vitek	Novel Therapeutic Reduces Abeta Deposition and Alzheimer's Pathology
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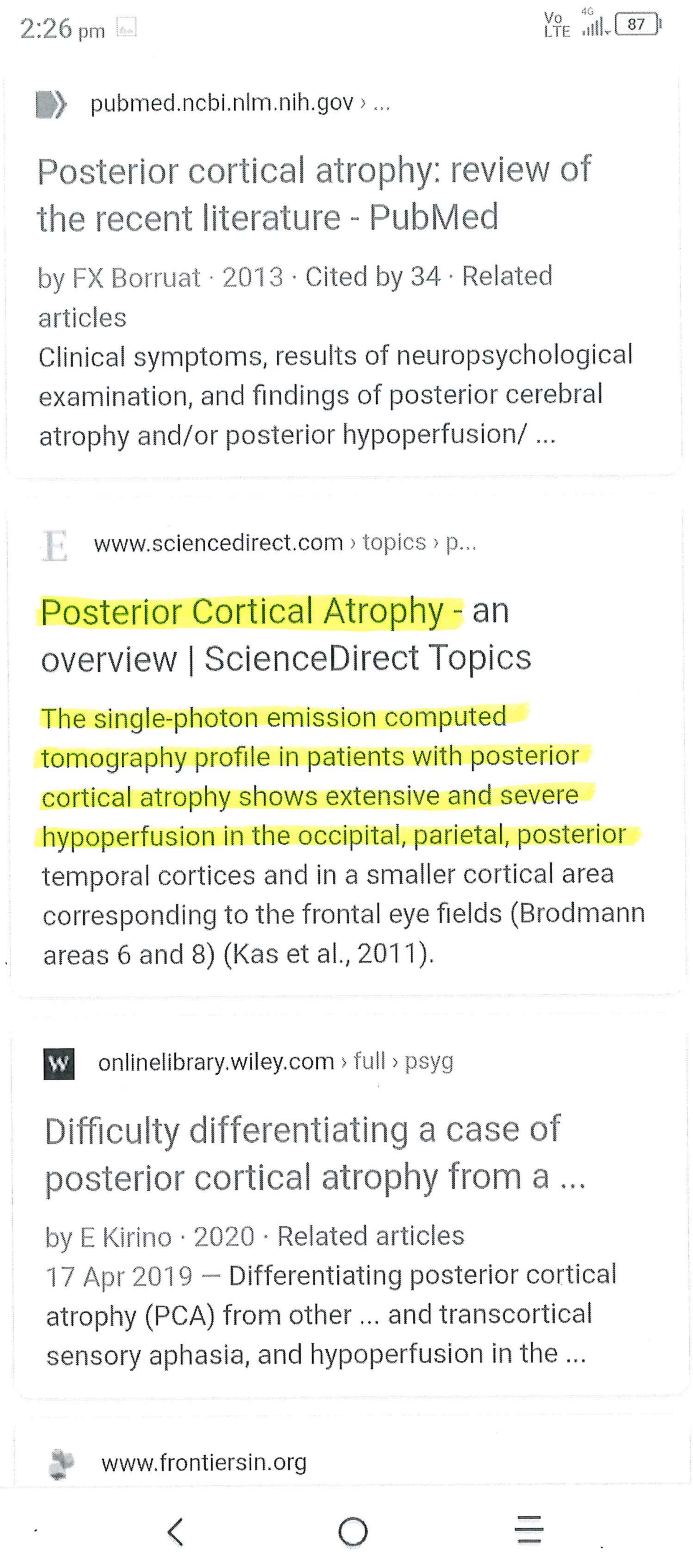

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Posterior cortical atrophy

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Posterior cortical atrophy (PCA) is a neurodegenerative syndrome that is characterised by progressive decline in visuospatial, visuoperceptual, literacy, and praxic skills. The progressive neurodegeneration affecting parietal, occipital, and occipitotemporal cortices that underlies PCA is attributable to Alzheimer's disease in most patients. However, alternative underlying causes, including dementia with Lewy bodies, corticobasal degeneration, and prion disease, have also been identified, and not all patients with PCA have atrophy on clinical imaging. This heterogeneity has led to inconsistencies in diagnosis and terminology and difficulties in comparing studies from different centres, and has restricted the generalisability of findings from clinical trials and investigations of factors that drive phenotypic variability. Important challenges remain, including the identification of factors associated not only with the selective vulnerability of posterior cortical regions but also with the young age of onset of PCA. Greater awareness of the syndrome and agreement over the correspondence between syndrome-level and disease-level classifications are needed to improve diagnostic accuracy, clinical management, and the design of research studies.

Introduction

Posterior cortical atrophy (PCA) is a neurodegenerative condition characterised by a progressive, often striking, and fairly selective decline in visual-processing skills and other functions that depend on parietal, occipital, and occipitotemporal regions of the brain. Age at onset of PCA is typically 50–65 years, and the syndrome is associated with various underlying pathological features. PCA has been recognised for more than two decades and yet, compared with other conditions, is relatively neglected by researchers. Patients often face considerable delays in diagnosis owing to the young age at onset and unusual symptoms at presentation. Furthermore, the term PCA has been applied inconsistently, making comparison across studies difficult to achieve. Although there have been moves to define neurodegenerative diseases by their underlying pathological features, such progress in relation to PCA is limited currently by scant specificity in available diagnostic criteria and an absence of clarity with respect to relations between PCA and related syndromic classifications, such as aphasic, amnestic, and dysexecutive Alzheimer's disease phenotypes and corticobasal syndrome.

In this Review we outline the clinical, psychological, imaging, epidemiological, genetic, and pathological features of PCA. We argue that, within pathological subgroups, characterisation of atypical phenotypes such as PCA will enable identification of biological factors that promote or protect against pathological changes in specific brain networks. Problems with and possible solutions to current diagnostic and terminological conundrums are considered, with particular reference to implications for future design of clinical and research trials that include people with PCA. We also aim to increase awareness and improve identification of early and unusual symptoms of PCA and to provide guidance on the provision of support, care, and education for patients, carers, and health-care professionals.

History and definitions

The term PCA was first used to describe people with predominant deficits in higher-order visual processing, a subset of whom also presented with striking atrophy in parieto-occipital areas of the brain.[1] The outlined syndrome accorded with other early reports of patients with similar clinical characteristics.[2–9] Without histopathological data, Benson and colleagues[1] deemed the clinical presentation of PCA to be sufficiently distinct from that of Alzheimer's or Pick's disease "to warrant classing them separately until definitive pathologic information becomes available". Subsequent histopathological studies identified Alzheimer's disease as the most common underlying pathological feature, leading to synonymous use of the terms PCA, biparietal Alzheimer's disease, and visual variant of Alzheimer's disease in some studies.[10–14] The term progressive posterior cortical dysfunction has also been used to describe clinical symptoms in affected individuals who do not have clear posterior atrophy.[15] However, PCA is also associated with pathological features that are not linked to Alzheimer's disease, which has led to calls for PCA to be regarded as a distinct nosological entity with its own diagnostic criteria.[16,17]

Epidemiology

The prevalence and incidence of PCA are currently unknown; obtaining these data will depend on the adoption of consistent diagnostic criteria. Furthermore, any value is likely to be an underestimate because of poor general awareness of the syndrome. However, Snowden and colleagues[18] noted that, of 523 patients with Alzheimer's disease presenting to one specialist centre for cognitive disorders, 24 (5%) had a visual presentation (also labelled PCA) and a further 13 (3%) had an apraxic presentation.

Age at onset tends to be much earlier in patients with PCA than in those with typical amnestic Alzheimer's disease, with most studies recording age of PCA symptom onset from mid-50s to early 60s,[17,19] although some researchers have reported a wider age spread (45–74 years,[20] 40–86 years).[21] In terms of sex distribution, no difference in prevalence has been noted on the basis of sex,[15,17,19] whereas other groups have reported an over-representation in women.[18,20–24]

Neuropsychological features

The neuropsychological deficits cited most frequently in individuals with PCA are visuospatial and visuoperceptual impairments, alexia, and features of Bálint's syndrome (simultanagnosia, oculomotor apraxia, optic ataxia, environmental agnosia) and Gerstmann's syndrome (acalculia, agraphia, finger agnosia, left–right disorientation; figure 1).[15,17,19–21,25–31] Deficits in working memory and limb apraxia have also been noted.[20] Findings of longitudinal studies indicate that anterograde memory, executive functions, and linguistic skills, which are sometimes preserved strikingly in earlier stages of disease, gradually deteriorate in some patients with PCA as they progress to a more global dementia state.[10,19,23]

Although higher-order visual problems, such as difficulties with object and space perception, are reported more often than are basic visual impairments, many such problems are at least partly due to deficits in more basic visual processing—eg, form, motion, colour, and point localisation. In a detailed comparison of basic and higher-order perception,[32] all patients with PCA showed impairment in at least one basic visual process, emphasising the vulnerability of fundamental aspects of vision associated with occipital cortical dysfunction. This study also indicated specific correlations of basic visual processing with higher-order visuospatial and visuoperceptual skills, but not with non-visual parietal functions (such as calculation and spelling), suggesting specific involvement of visual networks in PCA.[32]

Basic and higher-order visual deficits, in combination, have predictable outcomes on the performance of PCA patients on general neuropsychological tests—eg, performance IQ in patients is often up to 30–40 points lower than verbal IQ. Performance on cognitive tasks with any relevant visual component (eg, visual memory recall, trail-making test, Stroop test) is vulnerable to impairment and misinterpretation, and thus accurate assessment requires selection of tasks that minimise visual demands (eg, auditory-verbal memory tasks, naming from verbal description).

Many patients with PCA also have unusual symptoms, referred to as positive perceptual phenomena, which include abnormally prolonged colour after-images,[33] reverse size phenomena,[34,35] perception of movement of static stimuli,[35] and in one individual,[35] 180° upside-down reversal of vision. Reading skills can be limited by several processes, including visual disorientation (getting lost on the page), reverse size phenomena (accurately perceiving small but not large print), and visual crowding (impaired identification of constituent letters of a word owing to excessive integration of visual features from surrounding letters).[36–38] PCA can also lead to primary peripheral dyslexias.[39,40] Anecdotally, individuals with PCA frequently report heightened sensitivity to glare from shiny surfaces and can experience a range of localised sensation and pain phenomena and disturbances of balance and bodily orientation, which could potentially be linked to deranged visuovestibular interactions.

Should PCA be deemed a unitary clinicoanatomical syndrome or, rather, a collection of related but distinct syndromic subtypes? By extrapolation from basic neuroscientific evidence of distinct cortical streams that process different types of visual information,[41,42] researchers have suggested that separate parietal (dorsal), occipitotemporal (ventral), and primary visual (striate cortex, caudal) forms of PCA exist.[12,13] However, these claims are based on findings from individual case reports. Subsequent studies of neuropsychological case series have failed to provide evidence to support a pure ventral-stream syndrome[19] and, rather, have indicated considerable overlap in neuropsychological profiles and patterns of cortical thinning in patients with behaviourally defined predominantly dorsal-stream or ventral-stream

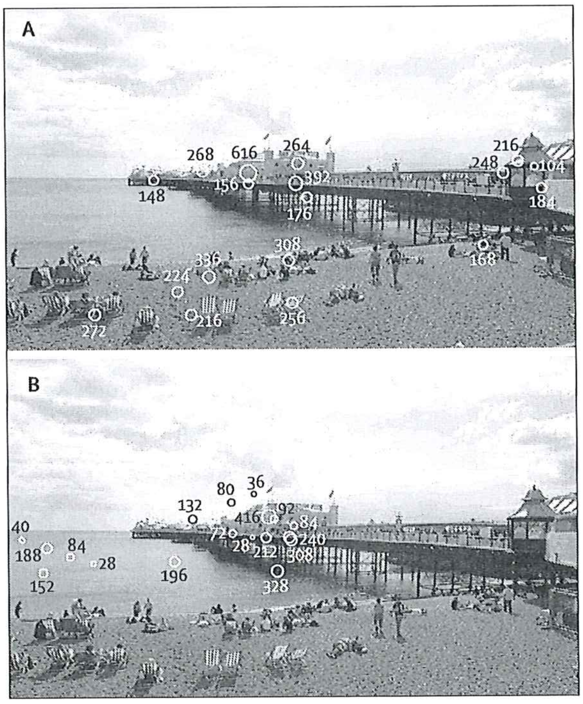

Figure 1: **Visual dysfunction in posterior cortical atrophy**
Individuals with posterior cortical atrophy have difficulty identifying objects and faces, particularly when they consist of many parts or are viewed from an unfamiliar (non-canonical) perspective. Eye-tracking studies contrasting scene perception in healthy individuals (A) and people with posterior cortical atrophy (B) suggest that patients have poor top-down guidance and control of oculomotor function. Circles represent fixation locations and circle size represents fixation duration. Patients with posterior cortical atrophy fixate prominent features initially (eg, dome on pier), but subsequently fixate relatively uninformative aspects of the scene (eg, sea or sky) and miss important contextual details (eg, beachfront or near the end of the pier). Images from Tim Shakespeare and Sebastian Crutch (unpublished).

impairments.[32] Thus, these findings suggest that phenotypic differences might most appropriately be judged to represent points on a continuum of variation within PCA.

Panel: Case study

A 62-year-old right-handed woman presented with 4 years of progressive visuospatial dysfunction. Her first symptom was difficulty seeing when driving at night. In the following years she frequently dented her car when parking, tended to bump into doors on her right side, and had trouble locating items even when they were directly in front of her. She reported problems reading, trouble distinguishing between currency bank notes, and difficulty deciding whether to push or pull a door to open it. When she watched television, images seemed to move slowly. She was referred to the cognitive neurology clinic by an ophthalmologist, who had ruled out primary ocular disease. On neurological assessment she was fully oriented and was an excellent historian. On testing of visual fields, she was inconsistent in counting fingers in the right hemifield. Pupillary responses and extraocular movements were normal, although she was slow initiating saccades and had difficulty reaching for items under visual guidance. Her physical neurological examination was otherwise normal. On cognitive testing, her MMSE score was 26/30, and she showed severe impairment when copying intersecting pentagons and the Benson figure (figure 2A). She was able to name colours correctly but showed moderate difficulty matching faces. She had severe difficulty reading, which was improved by spelling words out loud, and had mild deficits in confrontation naming (improved with cues) and category fluency. Verbal memory, phonemic fluency, and attention were intact. She could not complete many tasks because of visual dysfunction. Brain MRI showed striking atrophy in bilateral parietal, posterior temporal, and lateral occipital cortex (figure 2B [upper row]), and fluorodeoxyglucose (FDG)-PET (figure 2B [middle row]) showed hypometabolism in the same regions, left worse than right. Frontal cortex, medial temporal cortex, and hippocampus were spared. Pittsburgh compound (PiB)-PET showed diffuse cortical uptake throughout posterior and anterior cortical regions alike (figure 2B [lower row]), consistent with underlying amyloid-β plaques.

Clinical features

Clinical presentation of PCA is affected by several factors. These include time taken before an individual presents to medical services or is referred to a cognitive specialist; the specific pattern of deficits; in some people, the underlying pathological features; and the patient's psychological response to their symptoms. The relative rarity of PCA, the sometimes unusual nature of its symptoms, and the fairly young age at onset can lead to misdiagnosis of many patients as depressed, anxious, or even malingering in early stages of the disease. Early anxiety is (at least anecdotally) a common feature, perhaps indicating that patients with PCA typically have some insight into the possibility that they have a medical problem, even if its nature is unclear. Furthermore, even to experienced cognitive neurologists, a patient's initial history can be more suggestive of anxiety, until examination shows impairment that relates to function of the parietal lobe, the occipital lobe, or both of these areas. Patients are usually referred first to opticians and ophthalmologists in the belief that an ocular abnormality is causing their visual symptoms, sometimes leading to unnecessary medical procedures such as cataract surgery.

The symptoms reported by a patient with PCA probably indicate broadly their individual pattern of neuropsychological impairment. Visual symptoms are perhaps more likely to be mentioned than other posterior deficits, with individuals describing difficulties reading lines of text, judging distances (often leading to repeat minor car accidents or difficulties parking), identifying static objects within the visual field, or having problems with stairs and escalators (panel). Visual symptoms such as light sensitivity or visual distortions can be mistaken for migraine. Careful history taking could reveal some of the more unusual visual phenomena described above, including the presence of prolonged after-images or visual crowding. Individuals might volunteer difficulties in using common objects, suggesting dyspraxia, or describe progressive difficulty with calculations or spelling. The presence of other neurological symptoms, including visual hallucinations (reported in up to 25% of patients with PCA)[19,21,43] and rapid eye movement (REM) sleep behaviour disorder, could be suggestive of underlying dementia with Lewy bodies. Very occasionally, the patient's history might be consistent with occipital lobe seizures.

Careful bedside testing can elicit signs of disproportionate parietal or occipital dysfunction, including (but not restricted to) visual disorientation, difficulties resolving degraded stimuli, ideational or ideomotor dyspraxia (or both), dyscalculia, and problems with spelling (figure 2A). The physical examination in most cases of PCA is unremarkable; however, if severe cortical visual impairments are present, interpretation of visual acuity and visual fields can be difficult, with hemianopia sometimes misdiagnosed owing to the presence of higher-order visual attentional impairments. Finger myoclonus is also not uncommon. Snowden and colleagues[18] reported similar frequencies of extrapyramidal signs (41%), myoclonus (24%), and grasp reflex (26%) in individuals with PCA compared with those seen in patients with typical Alzheimer's disease. Nonetheless, features of clear symmetrical motor parkinsonism (suggestive of Lewy body pathology) or prominent asymmetric myoclonus and dystonia (suggestive of corticobasal degeneration) can give important clues to the underlying cause of PCA, although pathological data on which these clinical observations can be confirmed are currently sparse.

Neuroimaging

Increasingly advanced image analysis techniques have been used to localise and quantify (typically group) differences in patterns of atrophy in patients with PCA, compared either with controls or with patients with typical Alzheimer's disease. Cross-sectional voxel-based morphometry has shown widespread differences in grey matter between patients with PCA and healthy controls, with the most significant reductions found in regions of the occipital and parietal lobes followed by areas in the

Because of poor awareness of PCA, patients with this disorder typically receive scant or inappropriate care and advice, whereby difficulties that are less important to the individual (eg, memory problems) are targeted while vital perceptual difficulties (eg, many activities in day centres and nursing homes are visually mediated) are, in general, not considered. Preservation of skills such as memory, language, and insight in PCA, especially in mild-to-moderate stages of the disease, enables patients to take advantage of peer-support meetings and group, couple, and individual psychological treatments when the need exists. Support group meetings are especially useful in tackling social isolation, allowing patients to share the experience of what is often a long and difficult route to diagnosis, and to exchange practical tips and coping strategies and advice. Patients with PCA often benefit from resources designed primarily for blind and partially sighted people, such as mobile phones with simplified displays, voice recognition software, talking books and watches, culinary aids, and lamps to increase ambient light levels in the home. Referral to an occupational therapist or sensory team could be appropriate to help a patient participate more fully in activities of daily living. An individual might also need to be referred to an ophthalmologist to register as partially sighted under statutory invalidity schemes, which might then provide access to financial and social benefits and services. Driving a car is not appropriate for many people with PCA, particularly those with prominent visual disturbance. Physical therapy can also be helpful for individuals with parkinsonism and gait disturbance. Scant empirical evidence shows the effect of management strategies in PCA, but a rehabilitation programme that included psychoeducation, compensatory strategies, and cognitive exercises was tested in an individual with PCA,[79] resulting in small improvements in visuoperceptual functioning.

Conclusions

Posterior cortical atrophy is a debilitating and under-recognised focal degenerative syndrome that is associated with a range of different disease pathologies. The core features of the syndrome are sufficiently homogeneous to justify regarding PCA as an independent nosology, with Alzheimer's disease as the most common underlying cause. However a lack of consistency in the classification of PCA is likely to continue unless diagnostic criteria and terminology are standardised. The criteria proposed in this Review attempt to take both the clinical and histopathological features of PCA into account and to introduce quantifiable behavioural inclusion criteria for research studies of PCA. Better understanding and awareness of the syndrome by medical and lay workers is necessary to improve diagnosis and treatment and to enhance support services for individuals with PCA and their families. Identification of the distinctive patterns of structural, functional, cognitive, and genetic changes in PCA could provide new insights into the pathogenesis and clinical features of typical Alzheimer's disease and into general mechanisms of visual network function and degeneration. Dedicated trials are needed to assess the effectiveness of pharmacological and non-pharmacological interventions in PCA, and to identify factors that drive phenotypic variability in this small but important population of patients who typically have early-onset dementia.

Search strategy and selection criteria

We identified references for this Review by searching PubMed with the terms: "posterior cortical atrophy", "biparietal Alzheimer's disease", "visual dementia", and "Balint's syndrome dementia". We looked for articles published between 1970 and November, 2011. We also identified articles by searching our own files. Only reports published in English were reviewed, with the exception of historical manuscripts (before 1988). The final reference list was generated on the basis of originality and relevance to the broad scope of this Review.

Contributors

All authors contributed to the writing and reviewing of this Review.

Conflicts of interest

NCF has served on scientific advisory boards of the Alzheimer's Research Forum, the Alzheimer's Society, and the Alzheimer's Research Trust, and holds a patent for QA Box that might accrue revenue. In the past 5 years his research group has received payment for consultancy or for undertaking studies from Abbott Laboratories, Elan Pharmaceuticals, Eisai, Eli Lilly, GE Healthcare, IXICO, Lundbeck, Pfizer, Sanofi-Aventis, and Wyeth Pharmaceuticals. JMS has received payment for undertaking studies from AVID Radiopharmaceuticals. SJC, ML, GDR, and MNR declare that they have no conflicts of interest.

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Short Communication

Pronounced Impairment of Everyday Skills and Self-Care in Posterior Cortical Atrophy

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Abstract. Posterior cortical atrophy (PCA) is a neurodegenerative syndrome characterized by progressive visual dysfunction and parietal, occipital, and occipitotemporal atrophy. The aim of this study was to compare the impact of PCA and typical Alzheimer's disease (tAD) on everyday functional abilities and neuropsychiatric status. The Cambridge Behavioural Inventory-Revised was given to carers of 32 PCA and 71 tAD patients. PCA patients showed significantly greater impairment in everyday skills and self-care while the tAD group showed greater impairment in aspects of memory and orientation, and motivation. We suggest that PCA poses specific challenges for those caring for people affected by the condition.

Keywords: Caregivers, early onset Alzheimer's disease, psychology, questionnaires, self-care, self-report

INTRODUCTION

Posterior cortical atrophy (PCA) is a neurodegenerative syndrome characterized by progressive decline in visuospatial and visuoperceptual skills, literacy and praxis; with parietal, occipital, and occipitotemporal atrophy. Age of onset is typically in the 50 s or early 60 s; while the syndrome is most commonly caused by Alzheimer's disease (AD), cases due to dementia with Lewy bodies and corticobasal degeneration have been described [1, 2].

In contrast to the amnestic predominance of typical AD (tAD), presenting features in PCA commonly include difficulties with visually-guided behaviors such as driving, reading, or locating objects, while insight and episodic memory remain relatively spared [3]. It is likely that these cognitive impairments limit patients' ability to realize, adapt, and monitor interactions with their visual and physical environment. Our characterization of how PCA impacts upon everyday life is informed by clinical experience and patient anecdotes, but there has been little work to quantify or directly compare functional abilities between PCA and tAD.

The Cambridge Behavioural Inventory (CBI) is a carer questionnaire that evaluates a range of everyday skills and neuropsychiatric symptoms. Distinct profiles have been demonstrated for AD, behavioral-variant frontotemporal dementia, and Parkinson's disease [4]. There is high test-retest reliability [5], and a good correlation between its neuropsychiatric items and the Neuropsychiatric Inventory [6]. Since its inception, it has been revised (CBI-R) to reduce the number of items from 81 to 45, while retaining the most informative items for comparing tAD with frontotemporal dementia [7].

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Table 1
Individual items in the CBI-R in which there was a significant difference between the proportion of patients rated as showing a severe impairment in the PCA and tAD groups. No shading denotes PCA worse than tAD; grey shading denotes tAD worse than PCA

	CBI-R individual items	
Domain	Question	*p* value
Everyday skills	Has difficulties writing (letters, Christmas cards, lists, etc.)	<0.001
	Has difficulties using the telephone	0.002
	Has difficulties making a hot drink (e.g., tea/coffee)	0.003
	Has problems handling money or paying bills	0.007
	Has difficulties using electrical appliances	0.025
Self-care	Has difficulties dressing self	<0.001
	Has problems bathing or showering self	0.042
Sleep	Sleeps more by day than before (cat naps, etc.)	0.020
Eating habits	Table manners are declining, e.g., stuffing food into mouth	0.035
Memory and orientation	Has poor day-to-day memory	0.007
	Asks the same questions over and over again	0.014
Motivation	Fails to maintain motivation to keep in contact with friends or family	0.034

The analysis was repeated with groups matched for MMSE (PCA $n = 32$, MMSE = 16.44; tAD $n = 32$, MMSE = 17.31), yielding similar results. Differences in everyday skills ($p = 0.003$) and self-care ($p = 0.006$) remained significant. With the exception of sleep (PCA greater impairment than tAD in repeated analysis; $p = 0.006$), group differences in the remaining domains were not statistically significant consistent with the original analysis.

Association of CBI-R total scores with MMSE and disease duration

Greater CBI-R score was significantly correlated with lower MMSE in tAD ($R = -0.28$, $p = 0.02$) but not PCA ($R = 0.07$, $p = 0.70$). Neither tAD ($R = 0.19$, $p = 0.12$) or PCA patients ($R = 0.15$, $p = 0.41$) showed a significant association between disease duration and total CBI-R score.

DISCUSSION

This study demonstrates the ability of the CBI-R to differentiate between everyday functional abilities in PCA and tAD. PCA patients showed greater impairment in everyday skills and self-care, while tAD patients showed trends toward greater impairment in stereotypic and motor behaviors, and motivation. At the item level, PCA patients were more likely to have severe impairments in everyday skills and self-care but also in sleep (more sleep during the day) and eating habits (declining table manners), while a greater proportion of tAD patients had severe impairments in day-to-day memory, repetitive questioning, and motivation to maintain social contacts.

The majority of functional deficits highlighted in PCA could be related to visuospatial and visuoperceptual impairments (e.g., writing, using the telephone, making hot drinks) or posterior cortical-dependent impairments in calculation (handling money/paying bills) or praxis (using electrical appliances). The 'declining table manners' item was designed to capture behavioral disinhibition but may reflect practical, visual- and praxis-mediated eating changes (e.g., eating with fingers owing to problems manipulating cutlery and perceiving location of food on the plate). The only item on which PCA patients were rated worse than tAD patients without an obvious posterior cortical functional explanation was the apparent increase in sleep. More detailed analysis of sleep would be required to explore this finding. No patients suffered from obstructive sleep apnea; while it is possible that patients with dementia with Lewy bodies underlying PCA contribute to disruption of sleep, differences were observed only for excess sleep during the day, not disturbed sleep at night.

With regard to neuropsychiatric symptoms, the results point toward relatively spared motivation in PCA, at least in terms of motivation to maintain contact with friends and family. This may relate to what carers describe as a maintained sense of purpose. Differences in motivation between PCA and tAD have not been investigated previously, but patients with tAD have been noted to have high levels of apathy [11, 12] which may relate to the motivation domain in the current study [5]. Our results suggest that PCA patients may have relatively lower apathy, warranting assessment using more detailed measures.

It is notable that while tAD patients were more severely impaired on 2/8 memory and orientation items, the groups did not differ in the memory and

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Researchers Join to Draw Posterior Cortical Atrophy Out of Shadows

30 Jul 2012

You are a neurologist, and in comes a man in his fifties. He bumps into the doorframe before sitting down on the armrest of his chair, awkwardly trying to position his body in the seat. He can't find the glass of water you place in front of him. Then he engages you in a perfectly coherent conversation about current events. Does this person have Alzheimer's disease?

He may—sort of. But that's not nearly the whole answer. Indeed, according to research clinicians who particularly care about patients like him, it is time to raise the profile of his peculiar and poorly understood condition. Two days before the official start of the Alzheimer's Association International Conference held 14-19 July 2012 in Vancouver, Canada, **Sebastian Crutch**, **Jonathan Schott**, and **Nick Fox** of University College London, UK, together with **Gil Rabinovici** of the University of California, San Francisco, called the first in-person gathering of a newly formed working group on posterior cortical atrophy (PCA). The U.S. Alzheimer's Association and Alzheimer's Research UK supported the meeting. The gathering drew a quorum of a larger group that had been forming over e-mail in the months before. More than 30 research clinicians from 20 centers in Britain, Canada, Australia, France, Germany, Holland, Italy, Spain, and the U.S. decided to move past their own idiosyncratic ways of defining PCA and see if they can accomplish more by working together. Each participating center has one to five dozen cases in its care.

The group's to-do list is long. Besides pinning down how rare this neurodegenerative syndrome truly is, they aim to state a consensus about its core symptoms and boundaries to other diseases, and to develop diagnostic aids to be shared with the neurology community at large. They also agreed to pool samples for a genetic study of PCA's peculiar features, and to do the groundwork for therapeutic trials specifically for these patients. "PCA is under-recognized and hugely untapped for research. It is strikingly different from typical AD," said Schott.

PCA is a degenerative syndrome that affects the back of the brain. Some neurologists view PCA merely as atypical AD, where the disease's amyloid deposits are distributed similarly to typical AD but its neurofibrillary tangles form more focally in occipital regions. In practice, however, PCA is essentially a clinical syndrome based on a "gestalt," as neurologists like to call it, of a collection of problems centered around visuospatial processing rather than memory.

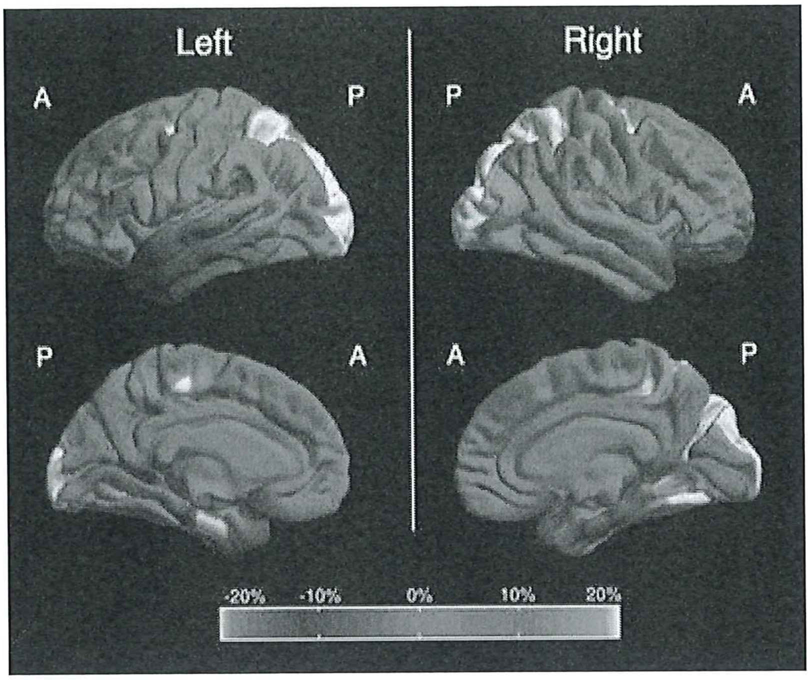
Many patients' initial symptoms largely fit that pattern. They cannot see objects they know are in front of them. They cannot construct in their mind's eye a map of where things are relative to each other. For example, blind people can use a mobile phone because they retain a mental image of the keypad, but people with PCA lose that internal spatial representation. Sometimes static objects to them seem to be moving. Some symptoms are counterintuitive. One patient in Fox's clinic, for example, recounted being unable to read the title of his daily newspaper when holding it in front of him on the train ride to work, while reading the small print just fine. Paradoxically, when looking down the train carriage, he could read the title on the same newspaper in the hands of a fellow commuter.

The symptoms can make patients disabled in daily life. Reading, writing, and calculation become difficult. People have to give up driving because they bump into things and misjudge the speed of other cars. They need help with mundane things such as finding light switches, putting a key into a lock, or picking out the can opener amid other gadgets in the drawer. "With a little help they can do almost everything; without help they can't do anything," Crutch recalled a carer saying.

At the same time, their memories are relatively intact, and most speak with acute insight into their condition. The British author Terry Pratchett, who eloquently raised awareness of AD through his own case, has a diagnosis of PCA. Even *The Man Who Mistook His Wife for a Hat*, from Oliver Sacks' famous book, has since been speculated to have suffered from PCA. If it is ever appropriate to compare devastating diseases, PCA may be the slightly lesser evil than typical AD, and Pratchett, for his part, has said as much. "In our support group, PCA carers feel like the patient is still the person they have always known," Crutch said.

Yet it's important for clinical centers to develop support tailored specifically to PCA. People with this disease feel poorly served by traditional AD support groups. That is partly because they are younger and face different challenges, and partly because the activities offered in AD day centers —puzzles, large-font books—are visual and thus "actively unhelpful for people with PCA," Crutch said. The UCL group runs regional support groups for some 200 patients with PCA in the UK and has a detailed website with information and patient videos.

The disease progresses slowly in some people and rapidly in others, but progress it does, and after some years, most patients do become memory-impaired and decline more broadly as their pathology spreads across the cortex.



This image shows in which regions of the brain the cortex is abnormally thin in people with PCA compared to people with AD. The color scale represents the magnitude of the difference in cortical thickness; left and right refer to the brain's hemispheres. Yellow and red represent more cortical thinning in PCA (occipital and parietal cortex); blue represents more cortical thinning in AD (mediotemporal, entorhinal, frontal region). *Summary image from 48 PCA and 30 AD cases, derived from Figure 1 in Lehmann et al., 2011.*

How common is PCA? Are the researchers starting up a big effort for a smattering of cases? Probably not, Fox said. Determining the true prevalence and incidence of PCA requires that consistent diagnostic criteria be in place. Specialist centers have reported that 5 to 10 percent of their referrals may fit the picture, more among early onset cases. Anecdotally, when a research proposal Fox's group had submitted to the UK Alzheimer's Society was being discussed in a meeting with caregivers, the question arose whether this was too rare to warrant funding. "Suddenly, every layperson in the room said, 'My husband has problems like that,' or 'The day center my wife goes to has someone like that,'" Fox said. From a pathology perspective, scientists know that some 25 percent of AD is atypical in terms of the distribution particularly of tangles. "We do not know what proportion of that is PCA. But even if it's 5 percent, that is a lot of people," Crutch said. In the U.S., that number would come to about 60,000.

That said, getting a PCA diagnosis can be a multiyear ordeal involving multiple visits to the ophthalmologist and prescription glasses until it's clear the problem does not lie with the eyes, said **Neil Graff-Radford** of the Mayo Clinic, Jacksonville, Florida. Primary care providers may not see a PCA case in years, and general neurologists dealing mainly with older patients may not always recognize it. Because the symptoms are so unusual, some patients have been dismissed as malingering, Crutch said.

POSTERIOR CORTICAL ATROPHY

YOUNGER ONSET DEMENTIA

Monitoring neuropsychiatric symptoms

Neuropsychiatric symptoms in PCA can contribute substantially to caregiver burden and distress; thus, assessing mood and behavioral changes through scales such as the Neuropsychiatric Inventory or Geriatric Depression Scale at regular intervals can be important for identifying appropriate psychopharmacologic and behavioral interventions for patients, and psychosocial support and education for caregivers.

Monitoring progression of general cognitive impairment: similarities/differences from typical Alzheimer's disease

Progression of cognitive impairments may be difficult to track depending on the subjective report of cognitive changes early in the course of the disease. Kennedy *et al.* [57] identified a healthy individual who presented with subjective memory complaints, but whose initial neuropsychological testing revealed deficits only in visual scanning (trail-making tests) and visual memory, and to a lesser extent, in object perception; verbal memory, visual reasoning and language were normal. Over the course of 4 years, deficits remained circumscribed to the visuospatial/visuoperceptual domain and continued to deteriorate, with the disorder progressing to include dyscalculia, dyslexia and dysgraphia; notably, performance on verbal memory measures remained stable and well within normal limits. This case highlights the importance of monitoring progression through serial neurocognitive investigation, in addition to attempting to obtain patient and informant reports of difficulties experienced in daily life.

With progression of the disease and development of dysfunction in cognitive domains other than visuospatial, patients with PCA increasingly resemble those with typical AD dementia albeit still with disproportionate impairment in the visuospatial domain. Compared with PCA patients, patients with typical AD dementia often show a far greater decline in short-term memory, executive functioning and language, with relative preservation of visuoperceptual abilities even well into the course of the disease. Comportment and behavior in typical AD may also deteriorate with disease progression. In the case of PCA, a patient can develop full-blown Balint or Gerstmann syndromes with progression of the disease, and experience a significant degree of visual impairment to be considered cortically blind. With a decline in sensorimotor function, PCA patients become increasingly dependent with respect to navigating their immediate environments and managing activities of daily living, including dressing and eating. Typical AD patients in more advanced stages usually retain procedural activities around dressing and eating, but may need increased cueing to complete the tasks due to lapses in memory or executive functioning.

Imaging & other biomarkers for monitoring progression of disease

We usually focus on repeating MRI scans (or CT with 3D reformatting in cases of MRI contraindications) to monitor progression of the brain disease causing a patient's PCA. Lehmann *et al.* [58] longitudinal investigation of atrophy patterns in PCA compared with tAD demonstrated the progression to a more diffuse pattern of atrophy from the initial, focal presentation. This pattern of widespread atrophy over the course of disease progression typically mirrors the deterioration in memory, language and executive functioning skills despite preservation in earlier stages of PCA. Although we are currently investigating the value of FDG-PET, amyloid PET, tau PET and CSF as longitudinal biomarkers of disease progression, we do not advocate for their use longitudinally in clinical practice. At some point, it may become clear that they are useful for monitoring progression and the effects of treatments if disease-modifying interventions become available.

Current & future treatments for posterior cortical atrophy

As is the case for all neurodegenerative dementias, once the diagnosis of PCA is made, the discussion must turn to the fact that, at present, there are no disease-modifying therapies and few symptomatic therapies for PCA, none with strong evidence for their efficacy. PCA patients can, however, be registered as legally blind/handicapped and access services and resources available for the blind or partially sighted despite demonstrating normal acuity as seen early in the course of the disease. In general, cognitive and behavioral symptoms of PCA can be addressed, optimally by a multidisciplinary team of specialists [59]. Treatment includes pharmacologic and nonpharmacologic management of symptoms, management of co-morbid conditions, psychosocial support, and education of the patient and family [60].

Pharmacological treatment

Currently, there is no medication known to be effective in stopping or slowing the progression of PCA, as is the case for all neurodegenerative dementias today. To date, there has been one clinical trial examining the efficacy of donepezil in treating symptoms of PCA [61], which unfortunately failed to demonstrate any impact on neuropsychological outcomes.

Treatment is typically symptom-focused, and usually includes cholinesterase inhibitors traditionally used in AD. These medications may help some symptoms including memory loss or attention and concentration. Trials of levodopa or carbidopa can be introduced to address movement or gait disturbances should they develop over the course of the disease.

Few studies focusing on the management of neuropsychiatric symptoms in PCA exist. The empirical treatment approach resembles that of other forms of dementia. Depression, anxiety and mild agitation may be responsive to treatment with serotonin reuptake inhibitors or other related medications [62]. Sleep disturbance is an important symptom to treat in patients experiencing it, and may be addressed with melatonin or trazodone. For severe agitation, a low-dose atypical antipsychotic (usually quetiapine) can be helpful [63] as can low-dose valproate [64], but the risk of increased all-cause mortality should be balanced against possible benefits [65]. Psychosis will often respond to low-dose atypical antipsychotics such as risperidone [66].

Rehabilitative treatment

Neuropsychological rehabilitative programs have been reported in single case studies in PCA (e.g., [67,68]) but with minimal or no lasting improvements. The current standard of care for PCA patients involves referral to an occupational therapist who works with the patient and caregiver on learning and implementing compensatory strategies that support activities of daily livings/instrumental activities of daily livings. Strategies focus on ways to negotiate the household, functioning in the community, and adapting the work environment.

The OT evaluation begins with an assessment of an individual's strengths and limitations while performing functional tasks. Through this assessment, a client-centered treatment plan that maximizes safety and independence for the patient is developed and executed with the patient and involving a caregiver. Standardized assessments may include tests such as the clock drawing test [69] the motor free visual perceptual test [70] and functional performance-based assessments such as the kettle test [71] and the multiple errands test [72]. In addition to standardized assessments, the OT evaluation obtains an occupational profile. This profile records what activities the patient has engaged in, the patient's values and what professional, familial and communal roles the patient has played. It identifies among these activities those which the patient is unable to do or perform at the level of independence they once did. Interventions then focus on training strategies to compensate for limitations. Strategies include using senses other than vision, promoting use of intact cognitive skills and implementing environmental modifications (e.g., high contrast colors to help with item identification, increasing ambient lighting, placing Velcro on doorknobs to identify important rooms such as the bathroom). Use of visual cues to aid visually guided navigation has also shown some promise as an environmental adaptation [73]. Yong *et al.* [74] have also identified potential interventions targeting reading through the strategy of restricting visual field (through single- vs double-word presentations), which minimized spatial and oculomotor demands of text reading, and an intervention that involved moving words into a fixation box, which assisted in localization of words within sentences, reducing the tendency for visual disorientation, excessive crowding and fixation instability.

Education is provided to the patient and caregiver to increase awareness of the patient's deficits and how they impact day-to-day functioning. Patients often benefit from having caregivers present during all or part of the evaluation and treatment to help ensure that strategies learned in therapy can be carried over into the home environment successfully [75,76].

Psychosocial treatment

Support groups for both patients and caregivers can be helpful for reducing isolation, validating individual experiences and assimilating coping strategies and advice generated by peers. Psychoeducation helps patients and families manage expectations of functioning in daily life and understand medical prognoses. Cognitive–behavioral therapy has also been found effective in addressing neuropsychiatric problems that often develop in PCA [43].

PCA patients may withdraw from activities due to visuoperceptual problems that limit safe and consistent participation. This behavior can exacerbate the negative psychological impact that often already accompanies the disorder. It is critical, therefore, to ensure that the patient remains active as much as his/her visuoperceptual abilities

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EXPERT

A Form of Dementia that Affects Vision: Posterior Cortical Atrophy

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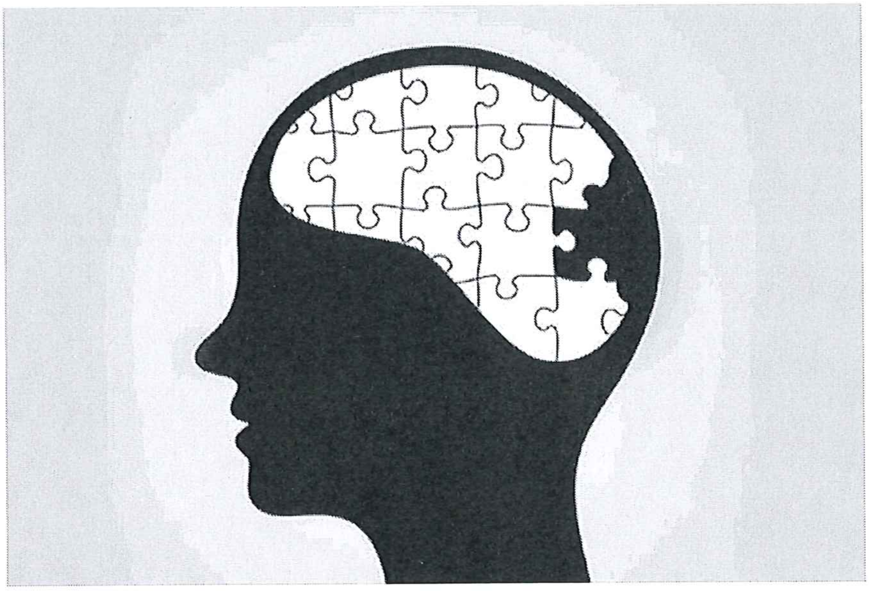

Learn about the symptoms, diagnosis, and treatment for a form of dementia where visual problems are a key feature of the disease.

"...She Doesn't See Them"

"Her car keys are right in front of her and she doesn't see them," her husband complained. "Maybe it's a blessing in disguise, because I'm worried about her driving too. She scraped the side of the garage twice last week when she was parking. She just got a new pair of glasses, but they haven't helped a bit."

Mrs. W* looked at the ground, embarrassed. She knew her husband's concern was a reflection of his love and frustration. At age 59, she thought she was young to have Alzheimer's disease (AD), but also felt certain that something was wrong with her thinking. She was getting more and more worried about this. Neuropsychiatric assessment found her memory to be only mildly impaired, but her ability to look at a cluttered picture and locate specific details was not as good as expected. She was sent for an MRI scan of the brain, which indicated some shrinkage in the back of the brain, called the **occipital lobes**. In that same brain region, her use of glucose, the brain's energy fuel, was reduced based on the results of a PET scan.

Posterior Cortical Atrophy

Mrs. W's condition is called posterior cortical atrophy (PCA). This form of dementia is easy to overlook. The key feature is usually visual problems without any discoverable eye disease, combined with cognitive problems that are milder than AD at the beginning. In Mrs. W's case, her most noticeable visual problem was difficulty "seeing" more than one thing at a time, a symptom that is called "simultanagnosia." In addition, she was younger than the typical AD patient and her memory and insight were only mildly diminished. She had none of the other findings or medical conditions, such as stroke, that might suggest a different diagnosis, and her imaging showed deterioration (atrophy) in the expected part of the brain.

PCA was first described in 1902, by the neurologist Arnold Pick. Since then, others have added to our knowledge, but we know much less about PCA than AD because PCA is not common. It is estimated to be present in about one of every twenty patients with dementia.

Symptoms

Patients with PCA experience the onset of symptoms earlier than those with AD, often in their mid-50s to early 60s. Memory and insight can be relatively good during the early stages of the disease, so patients often present with visual concerns. The damage with PCA is especially severe in the posterior part of the brain, the occipital lobes, which is why the disease is called posterior cortical atrophy. The occipital lobes help us understand what we see, so disease in that area creates visual problems. With PCA, a person may have trouble grasping the "big picture" or picking out something specific from a clutter of other objects. Things don't look right, so the patient may first turn to an eye doctor, but examining the eyes or changing lenses doesn't address the underlying brain problem.

In addition to visual and memory changes, PCA patients can show significant behavioral changes. Anxiety and irritability are the most common. Apathy or depression occur about as often as with AD. If the use of antipsychotics is considered as a treatment for hallucinations or other symptoms, care must be taken to recognize the risks of this class of medications use with dementia patients in general and DLB patients in particular.

Because of its early onset, PCA affects people still busily involved in their careers and even parenting responsibilities. The frustration of not being able to do things like driving, anxiety over loss of independence, and fear for the future can be very intense and depressing.

Diagnosis

Looked at through the microscope, PCA is often determined to be a form of AD. Autopsy of PCA patients show plaques and tangles in the occipital lobes and other brain areas typical for AD. Sometimes, though, the syndrome is an early version of other dementias such as dementia with Lewy bodies (DLB) or corticobasal degeneration. This may not be clinically apparent, though the presence of hallucinations makes it very possible that the cause is DLB rather than AD.

Brain imaging is useful in establishing the diagnosis of PCA. Early on, an MRI will typically show atrophy in the posterior part of the brain that is out of proportion to atrophy in other parts of the brain. FDG-PET, a measure of brain metabolism, will show reduced activity in the same brain region. In the PCA cases due to AD, PET-amyloid scans will show accumulation of amyloid in the posterior portion of the brain as well.

Treatment and Other Helpful Guidelines

As with AD, caregiver burden is difficult for those close to people with PCA. Because of its early onset, PCA imposes special and unique stresses. In addition, since PCA is uncommon, Mr. W was unable to find a support group in his area, but it was recommended that he join a group for early-onset AD, and he found a lot of comfort and practical advice by speaking with other caregivers there.

Summary

PCA is less common than AD, but it's important to consider it as a cause of visual and cognitive symptoms in a relatively young person. With correct diagnosis, a treatment team can provide explanation and support that takes into account PCA's special features. The care of a person with PCA requires careful attention to safety considerations, lifestyle changes, and planning for the future. PCA is not yet a curable condition, but an informed treatment team can lighten the burden for patient and caregivers.

*To protect anonymity and to illustrate features of PCA, Mrs. W's story is a composite of symptoms from various patients.

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When Glasses Don't Always Work … Posterior Cortical Atrophy and How Vision Changes with Dementia

COVID-19 Update: Click here for more information on senior safety.

Memory loss isn't the only common symptom for those with dementia. A senior with dementia can also experience vision impairment. Losing sight can be confusing, disorienting, and frightening to a senior who may respond with strange reactions. Family caregivers may mistakenly think the senior may be having visual hallucinations.

To better understand visual impairment, try holding your hand up in front of one eye. Unlike affected seniors, you will know the cause of your partial vision impairment and can easily remove your hand and see clearly again.

Defining Vision Loss: Posterior Cortical Atrophy

The medical term for reduced vision due to dementia is Posterior Cortical Atrophy (PCA). To better explain this term, let's break it up and define each separate word:

"Posterior" means the back part of the brain

"Cortical" means the thinking outer layer of the brain

"Atrophy" means shrinkage of that area of the brain

Atrophy is visible on CT scan and MRI scan

The complete term is confusing for and not easily remembered by family caregivers without professional healthcare experience. An easier term for this condition is PCA (not to be

Box 2. Posterior cortical atrophy: a caregiver's perspective.

"In my wife Judy's case, there were no bright line markers for the onset of PCA. Instead, we spent 5 or 6 years trying to get a proper eyeglass prescription starting in 2004. Her complaint was that she could not see, but she could pass the eye test at 20:20 with glasses. In addition, there were increasing episodes of forgetfulness and depression. Her driving was impaired. In retrospect, my wife clearly knew that something was not right and she very cleverly hid the symptoms, as best she could from me and herself. Our daughter Marie, living in California, clearly saw her mothers' decline and insisted that we do something more to help her mother. In October of 2007, the three of us traveled to Jacksonville, Florida, for appointments at the Mayo Clinic. As a result of four days of exams, we learned that Judy's eye problem was not her eyes but her brain. Their diagnosis was that Judy had PCA, a diagnosis that had not been on any of our radar screens. It was a relief to have a diagnosis, even though the prognosis was unfavorable. Judy bravely confronted her diagnosis. She was an evangelist for the local Alzheimer's Association. She described early-onset dementia to a number of audiences. She continued her daily walks with her dog and healthy eating habits. We continued her care at home, until it was too much. She moved into a memory care unit in March of 2011 where she died in March of 2018. As caregivers, we tried to provide for her in the best possible way."
– Fontaine Richardson

PCA: Posterior cortical atrophy.

and physical environment permits. Continued participation in meaningful activities is one way to avoid exacerbation of depression and anxiety. We typically encourage caregivers to identify and engage patients in structured and regular activities, such as those provided in day programs. Given the impediments to participating in visually mediated activities at day programs, incorporating therapies that involve use of other sensory modalities (as is the case in reminiscence therapy) may be particularly important to integrate into activities for PCA patients. Reminiscence therapy [77] can be helpful for stimulating recall of remote personal events, and will be particularly helpful once memory impairments become prominent in the PCA profile. PCA patients may also be well suited for music-based interventions involving exposure to familiar music or singing, which have become increasingly popular and being included in day-program activities.

Patient engagement in care planning

When a diagnosis of PCA is first made, many patients are still able to participate in care-planning activities, as reasoning, judgment, memory and communication skills are relatively preserved early in the course of the disorder. Although most patients appear to retain a fair degree of independence having spontaneously implemented compensatory strategies for reading, writing, navigation and finding personal effects, they have nonetheless begun to have difficulty functioning at their previous levels. We believe it is essential to engage each patient and family member in these discussions soon after diagnosis, typically focusing on adapting to the cognitive, physical and emotional challenges associated with PCA, and how best to plan for the coming years that will likely involve worsening of existing symptoms. Identifying new or worsening symptoms as well as areas of stability at each clinic follow-up are crucial in ongoing planning and care management. At some point, it may be helpful to 'step back' and engage in discussions of longer-term care planning while the patient is able to participate. The clinician(s) should clarify the patient's and family's knowledge of, experience with, and preferences regarding dementia care planning. In some cases, discussions may be conducted in the context of patient/family meetings. In other cases, it may be optimal to have these conversations with the patient separate from the family or other caregivers. The involvement of an experienced clinician (e.g., clinical social worker or occupational therapist) with dementia care planning skills is invaluable for these purposes. The Alzheimer's Association may offer local resources to assist with this fundamentally important process.

Neuropathology & genetics of posterior cortical atrophy

The neuropathology of PCA is usually AD. However, there are barely any clinicopathologic studies of PCA with more than five cases. Available studies suggest that AD neuropathology may account for 65 [6], 77 [5], to 100% [10] of cases. PCA may also be caused by pathology associated with corticobasal degeneration, Lewy body disease or rarely other neurodegenerative diseases [5]. We previously reported a case of frontotemporal lobar degeneration pathology presenting as PCA [11]. In patients with PCA due to AD, it is not yet clear why the localization of neuropathology is different than the typical localization in AD.

to identify individual fingers during the neurologic examination), and phenomena that can be both described as a problem by the patient and observed on formal testing (e.g., difficulty seeing more than one object at a time on a cluttered surface). To the best of our knowledge, there are no published instruments yet that support the assessment of PCA symptoms, and signs of the syndrome often are evaluated qualitatively rather than quantitatively. Here, we summarize the symptoms and signs that are currently used to diagnose PCA, acknowledging that the conceptualization of some of these phenomena is rooted in traditions of cognitive neurology and neuropsychology and may be in need of revision as relevant fields of cognitive neuroscience advance.

Typically, the initial clinical presentation involves report of an insidious onset and gradual progression of predominantly visuospatial or visuoperceptual symptoms; all other cognitive and behavioral domains are relatively spared in comparison, although not always entirely normal. Patients may describe visual disturbances such as blurred or partially obscured vision ('looking through a gossamer or veil') or glare sensitivity that interferes with reading, writing or negotiating the physical environment (e.g., resulting in tripping or mis-stepping off a curb, problems navigating and orienting geographically). In the face of normal ophthalmologic findings, further probing into the symptoms reveals impairments of higher-order visual processing with or without hemianopsia or other primary cortical visual loss. For example, patients may report difficulty reading digital clocks or signage. They are unable to identify or find objects in plain sight, especially when those objects are embedded in complex, cluttered environments (e.g., keys on a desktop or in a kitchen drawer); this is a condition known as simultanagnosia. Patients may also be unable to see objects on similarly colored backgrounds (e.g., a brown purse on a brown chair) or misjudge the placement of objects (e.g., a glass too close to the edge of a table). They often misjudge distance while driving, leading to minor scrapes and dents in an automobile, or bicycle or automobile accidents. Some patients report disorientation in the environment and getting lost in familiar areas (an example of topographical disorientation, where one loses sense of direction or ability to use landmarks for navigation). Visuoperceptual, spatial or higher level somatosensory or motor control disturbances that often occur in PCA can cause problems with dressing due to difficulty orienting oneself in relation to clothing (dressing apraxia), and problems reaching accurately for objects in space. In some cases, patients with PCA have difficulty recognizing familiar objects (visual agnosia) or familiar faces (prosopagnosia).

Patients with PCA can also present with elements of Balint syndrome. While the full-blown syndrome may not be present early in the course of the disorder, patients may report symptoms of what is ultimately determined on evaluation to be signs of simultanagnosia, optic ataxia and oculomotor apraxia – the core elements of Balint syndrome. These signs are typically ascertained on formal neurologic or neuropsychological examination, although experienced clinicians can recognize signs based on patients' descriptions of symptoms alone. For example, in the case of simultanagnosia, patients may report perceiving only parts of what is around them in a room or a scene, resulting in an inability to appreciate the overall meaning of what they see. They may be unable to attend to more than one object in view at a time, as evidenced by their description of only some of the objects or people in a room. They may also fail to direct visual attention and gaze in order to scan the visual field fully or respond to stimuli (also known as oculomotor apraxia). Finally, they may report 'clumsiness', or difficulty reaching for objects precisely (a symptom associated with optic ataxia).

PCA patients can also present with symptoms of Gerstmann syndrome. Although its validity as a distinct syndrome (rather than an artifact of observer bias, a reflection of aphasia or the result of extensive neurodegeneration) is debated [19], this syndrome includes acalculia, right–left disorientation (confusion with body-centered right and left side of space), agraphia, and finger agnosia. Acalculia may manifest as an impaired ability to perform simple calculations or a loss of numeracy (the appreciation of number values and magnitude, and/or the ability to reason and apply simple numerical concepts). Difficulty with calculations may occur also because of reading or writing impairment or disordered spatial organization of numbers on a page due to neglect, misalignment or number inversions. In these cases, patients report difficulties balancing their checkbooks, paying bills, transcribing numbers for addresses, counting out money for purchases or understanding measurements (e.g., for a recipe). Agraphia in these patients typically reflects visuoperceptual and visuospatial problems that result in an inability to write in a straight line or with normal spacing of letters and words, to copy the written word, or to sequence and orient writing properly. In a subset of cases, agraphia may also reflect spelling problems that result from a loss of orthographic knowledge. Finger agnosia typically is not apparent as it has less impact on daily functioning, as dexterity is relatively normal and does not manifest unless tested directly.

A dyslexic syndrome often develops in PCA, reflecting an inability to recognize the printed word or to generate eye movements/saccades from word to word such that they are able to track text on a page or move their gaze from the end of a printed line to the beginning of the next line. Patients may also experience more peripheral reading problems with visually complex text (e.g., handwritten or cursive text). Mendez *et al.* [20] found that PCA patients can develop an 'apperceptive alexia', resulting from the visuoperceptual and visuospatial deficits that lead to difficulty integrating text. The authors also suggested that phonological processing dysfunction may be present and contribute to reading difficulties.

A subset of PCA patients may also have word retrieval (due to anomia or naming problems) and comprehension difficulties, most noticeably in conversation. They may ask for questions or comments to be repeated, a behavior which appears to an outsider as a hearing difficulty or memory problem rather than a language difficulty *per se*. Such language impairment is consistent with findings by Crutch *et al.*, who found that PCA patients show disturbance of phonological processing and word retrieval/access [21]. Together, the symptoms of alexia, agraphia, and diminished comprehension and naming problems in the context of normal repetition and speech production constitute what Benson *et al.* [1] referred to as a transcortical sensory aphasia syndrome.

Praxis, or the ability to perform skilled, voluntary movements, often is affected in PCA as well. Patients describe problems with hand–eye coordination, but they or informants are rarely aware of this impairment unless it is severe enough to impact functioning in daily life. Apraxia is apparent with tool use or manipulation of common objects and most noticeable when a subject is asked to pantomime their use (e.g., striking a match and blowing it out, brushing one's teeth). In these situations, patients may use body parts as a stand-in for the objects rather than manipulate an imaginary object, suggesting they either do not understand that they are to pantomime or they cannot complete the task even though they understand it. Should patients still demonstrate difficulty pantomiming the action correctly despite feedback and the examiner's efforts to correct the patient (and in the absence of a language comprehension deficit), this is likely apraxia.

Are there subtypes of posterior cortical atrophy?

There is currently limited evidence from structural neuroimaging or neuropsychological studies to suggest discrete PCA subtypes. However, the following qualitative descriptions of different phenotypes have been suggested by Crutch *et al.* [9] as a starting point for research studies.

The biparietal (dorsal) variant is defined by the presence of "early, predominant, progressive visuospatial dysfunction, with features of Gerstmann or Balint syndromes, neglect and limb-kinetic apraxia." The occipitotemporal (ventral) variant is characterized by the presence of a predominant and progressive impairment of visual identification of objects, symbols, words or faces; this variant may include patients who report symptoms of a progressive prosopagnosia. In our experience, most of these patients also exhibit symptoms or signs suggestive of dorsal system involvement. The primary visual (caudal) variant of PCA involves mainly occipital cortical degeneration with predominant impairments in basic visual perceptual abilities; however, this variant is rare relative to the first two variants. Finally, a dominant parietal variant is associated with primary impairments in calculation, praxis and spelling, and arises as a result of more prominent involvement of dominant/left posterior cortices. This variant appears not to be rare in the PCA spectrum and may in fact be a 'non-visual' presentation of the disorder with elements of Gerstmann syndrome and transcortical sensory aphasia. Notably, this presentation is not to be mistaken with disorders such as logopenic-variant primary progressive aphasia (lv-PPA): in the case of PPA, detailed investigation may reveal impairments in visual cognition, but they are not the primary symptoms or signs of the disorder.

What differentiates posterior cortical atrophy from a typical Alzheimner's disease clinical syndrome?

In contrast to PCA, typical AD dementia is characterized by initial presentation and early dysfunction in episodic memory. Visuospatial deficits occur at the stage of mild dementia, but at a level less severe than those observed in the early stages of PCA. The visuospatial deficits in typical AD are relatively mild in comparison to other symptoms and signs, and manifest on visual construction tasks or in activities reliant on spatial function (i.e., navigation). More complex deficits such as simultanagnosia and visual hemineglect do not usually occur until the moderate stage of typical AD dementia.

Onset of episodic memory dysfunction in PCA is variable and a matter of some debate and active research (e.g., [22]). In our experience there is substantial variability in the types of memory problems demonstrated in PCA. Patients may report forgetfulness for conversation details or recent events, misplacement of personal effects or describe 'action lapses', where they enter another room and momentarily forget the reason. These lapses, however, can

Posterior Cortical Atrophy: A Difficult Diagnosis

Erin Draper

Abstract

Background: Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative condition with initial symptoms of subjective visual loss. Affected patients report difficulty reading and other visuoperceptual and visuospatial impairments. Neuro-imaging of PCA patients demonstrates increased atrophy of the parieto-occipital cortex, consistent with corresponding cortical visual dysfunction.

Case Report: A 70 year old woman presented for a low vision evaluation secondary to her complaint of inability to read of two years duration. Cataract surgery yielded no improvement. Previous retinal examination and report of neuroimaging was normal. Her best corrected single letter distance acuity measured 20/40 and her best corrected single letter near acuity measured 20/50, however, she was unable to read words regardless of the enlargement ratio. With confrontational field testing she exhibited severe general constriction, despite full functional awareness. There was no central scotoma noted. A multifocal electroretinogram was completed and normal. A mini-mental status exam revealed severe cognitive impairment. A review of prior imaging uncovered pathologic posterior cortical atrophy. Upon questioning, her family reported memory decline, prosopagnosia and issues with visuospatial perception. The diagnosis of posterior cortical atrophy was made.

Conclusion: Posterior cortical atrophy is a visual variant of Alzheimer's disease. It is under-recognized and often when it is, has a delayed diagnosis. Initial symptoms are typically visual-related; patients present with a normal ocular health exam but symptoms indicate impaired higher order visual processing. Neuro-imaging and neuro-psychological testing are key components of diagnosis. Low vision rehabilitation with text-to-voice technology and simplified video displays can improve quality of life. We review PCA, its epidemiology, signs and symptoms, differential diagnoses, work up and low vision rehabilitative solutions.

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Journal of Geriatric Psychiatry and Neurology

The Difficult Diagnosis of Posterior Cortical Atrophy in a 62-Year-Old Woman

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Abstract

Posterior cortical atrophy (PCA) describes a rare heterogenous neurodegenerative syndrome with early visuospatial and visuoperceptual deficits due to atrophy of parieto-occipital brain regions. Here, we describe the case of a 62-year-old woman showing severe cognitive impairments as well as hemianopsia and all core symptoms of Bálint's syndrome. Years ago, the patient had complained about a "tunnel view" and concentration problems. The diagnostic results point to a case of PCA with underlying Alzheimer pathology. The disease course until diagnosis lasted for 7 years, reflecting the diagnostic difficulties with this still largely unknown syndrome. The unfamiliar symptom presentation including fluctuations in cognitive performance, affective symptoms, cerebrospinal fluid (CSF) biomarkers, which were at first inconspicuous, and a former suspected diagnosis of dissociative pseudodementia, altogether brought considerable uncertainty to the involved health-care professionals. We conclude that cases of "atypical dementia" presenting with visual symptoms, even if appearing unspecific at first, are suspect of PCA. This case report provides an ostensive overview of PCA, including imaging data, CSF-findings, original drawings and handwriting samples from the patient.

Keywords

posterior cortical atrophy, Alzheimer's disease, Bálint's syndrome, hemianopsia

Background

Posterior cortical atrophy (PCA) is a rare neurodegenerative syndrome with an early age of onset in the mid 50s to early 60s. Initially described as visual Alzheimer variant, the underlying pathology, however, in some cases, can also be attributable to other neurodegenerative entities.[1] The pattern of cortical atrophy shows an early accentuation of parieto-occipital brain regions involving important fields of cortical visual representation, whereas areas affected early on in Alzheimer disease (AD), like the middle temporal lobes, often are spared for a relatively long period.[2,3] Accordingly, the first symptoms predominantly concern visuospatial and visuoperceptual functions, whereas higher mnestic functions are normally affected later in disease course.[4,5] Patients often have a good insight into their symptoms, whereas AD often is associated with anosognosia.[6] Together with the logopenic variant of primary progressive aphasia and early-onset AD, PCA might be part of a clinical continuum of early-onset dementias that share clinical and neuroanatomical features.[7] The consensus criteria from 2017 suggest a 3-step diagnostic approach with diagnosis of the clinical–radiological syndrome requiring at least 3 core symptoms of visuospatial or visuoperceptual impairment at disease onset in contrast to relatively spared memory, executive, and speech functions as well as preserved personality and behavior.[8] While these criteria discriminate against AD, as well as language and behavioral variants of frontotemporal lobar degeneration, additional exclusion criteria account for possible somatic causes like strokes, lesions of the visual pathway, or brain tumors. The radiological signs, however, are facultative, which makes PCA an all-clinical diagnosis. When clinical criteria are met, in a second step it shall be checked if the disease is associated with a neurodegenerative syndrome other than AD (ie, corticobasal syndrome, Lewy body dementia, or prion disease). Finally, the underlying pathology shall be clarified by biomarker analysis in cerebrospinal fluid (CSF), which nowadays is only possible for Alzheimer pathology (Aβ 1-42, Tau, p-Tau) and prion disease (14-3-3 protein, RT-QuIC method).

Case Description

A 62-year-old married woman was admitted to our Neurogerontopsychiatric Day Care Unit due to cognitive deficits. The first abnormalities dated back 7 years, when the patient still worked as a physiotherapist, but developed problems with reading and writing professional letters as well as with performing simple calculations. Due to these issues, the patient had to quit her job 1 year later. Five years ago, the patient complained about a “tunnel view” which led to repeated consultations of opticians to adjust new glasses without any contacts to ophthalmologists. Spatial disorientation, being helpless in unknown places, and not being able to drive any longer occurred 3 years later. For 2 years now, the patient has disorientation in time, and daily life has been impaired by loss of practical abilities, like handling the cooker, the washing machine, the telephone, or the TV remote control. Today, the patient needs personal assistance for dressing (wears clothes inside out or is not able to put them on) and eating (leaves food on the plate). Also, difficulties in grasping things like cutlery are described. Concentration problems were accompanying these symptoms, severe memory deficits, however, being described for the last 2 years only. Symptoms and results of cognitive screening tests are listed in Table 1. The first professional appointment was 5 years ago, when the patient was examined in a neurologic hospital, which resulted in a suspected diagnosis of beginning dementia of unclear entity and prescription of donepezil. The CSF biomarkers were normal then (Table 2). During another hospital stay half a year later, the amnestic deficits were diagnosed as mild cognitive impairment which was attributed to an “atypical depressive syndrome” and even a dissociative etiology has been discussed due to the symptom presentation leaving a “demonstrative” impression and a long-lasting marital conflict as a potential psychogenic factor. In the mini-mental state examination (MMSE), the patient scored 24/30 points then, remarkably with the deficits occurring in the visuoconstructive (−1) and the calculation (−5) task without memory impairment (Table 1). Neurologic examinations had been described as unremarkable so far. The medical record showed a history of arterial hypertension and Hashimoto’s thyroiditis. There were no relevant childhood diseases or pre/perinatal complications. Twice, at the age of 26 (postpartum) and 34, the patient had an acute polymorphic psychosis. Both episodes remitted after a few days of antipsychotic medication with no need of medicamentous recidive prophylaxis. The patient had no history of alcohol or drug abuse. The family history was empty of neuropsychiatric disorders.

Click to view table

Table 1. Symptoms and Results of Cognitive Screening Tests in 2013 and 2018.[a]

Click to view table

Table 2. Cerebrospinal Fluid Findings.[a]

Investigations

In our examination, the patient was disoriented in time, place, and situation. Concentration and short- and long-term memory were severely impaired. The patient could not subtract 7 from 100, could not spell short words, could not recall any of 3 words after 2 minutes, and had difficulties remembering the names of her 2 sons. Severe word finding difficulties appeared. Apart from that, the speech was fluent with no signs of agrammatism, semantic, or phonemic paraphasia. Insight into the deficits was preserved by the patient, being described as feeling of an "empty head with nothing left inside." The overall mood was good, the affect, however, being slightly labile, quickly responding to both positive and negative emotional stimulation. Additionally, the patient was very irritable in new situations, then she sometimes reacted short-tempered and seemed to be helpless. Formal thinking was a bit slow but sorted. There were no delusions and no hallucinations. The neurologic examination showed hemianopsia to the right. Eye saccades were uncontrolled and apractic, but there was no gaze palsy. Otherwise, the brain nerves were inconspicuous. The right-handed patient had difficulties imitating hand movements with no side preference as a sign of ideomotoric apraxia. The description of Binet's images and Navon figures was hampered due to simultagnosia. The clock drawing test[10] as well as free drawing attempts showed severe visuospatial deficits (Figure 1). The patient had agraphia (Figure 2) and alexia. The drawing of complex figures was disabled (Figure 3). Furthermore, we found an optic ataxia on the right side (atactic grasping of objects when coming into the visual field from the right). Apart from that, there were no focal neurologic defects, particularly no Parkinsonism and postural stability was fine. The neuropsychologic screening tests pointed to severe cognitive impairments (mini-mental state examination [MMSE] 9 points, montreal cognitive assessment test [MoCA] 4 points). The MRI brain scans showed a ballooning of the ventricular system, which was most pronounced in the left posterior horn with strong atrophy of left parieto-occipital cortices (Figure 4). In the fluorodeoxyglucose-positron emission tomography (FDG-PET) investigation a generalized cerebral hypometabolism was evident, most distinctive in left parieto-occipital cortex regions (Figure 5). The electroencephalography recording showed a permanent slowing with generalized theta, sometimes also delta-activity without any epileptic potentials or focal abnormalities. The CSF showed no signs of central nervous system inflammation or blood–brain barrier impairment. Tau and Phospho-Tau protein were normal, β-amyloid 1-42 was markedly decreased (Table 1) at the current investigation, whereas in 2013, it still was above cutoff. The results of the routine blood test were inconspicuous.

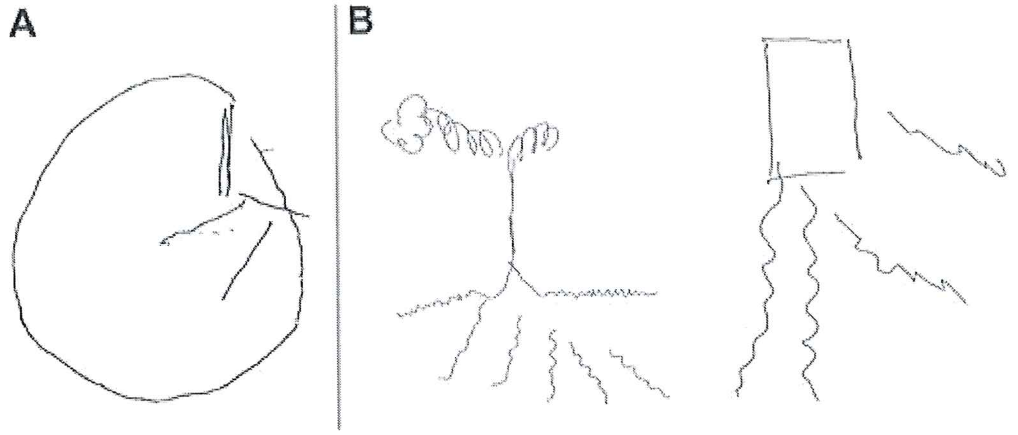

Figure 1. Visuospatial disorganization. Clock drawing (A): Task to draw the conture, all numbers, and the hands at 10 past 11. The performance hints to severe visuospatial disorganization (score = 5). Free drawing attempts (B), a tree, and a house, are 2-dimensional, poor in details, and leave some sections unconnected to the rest of the image illustrating the patient's visuospatial deficits.

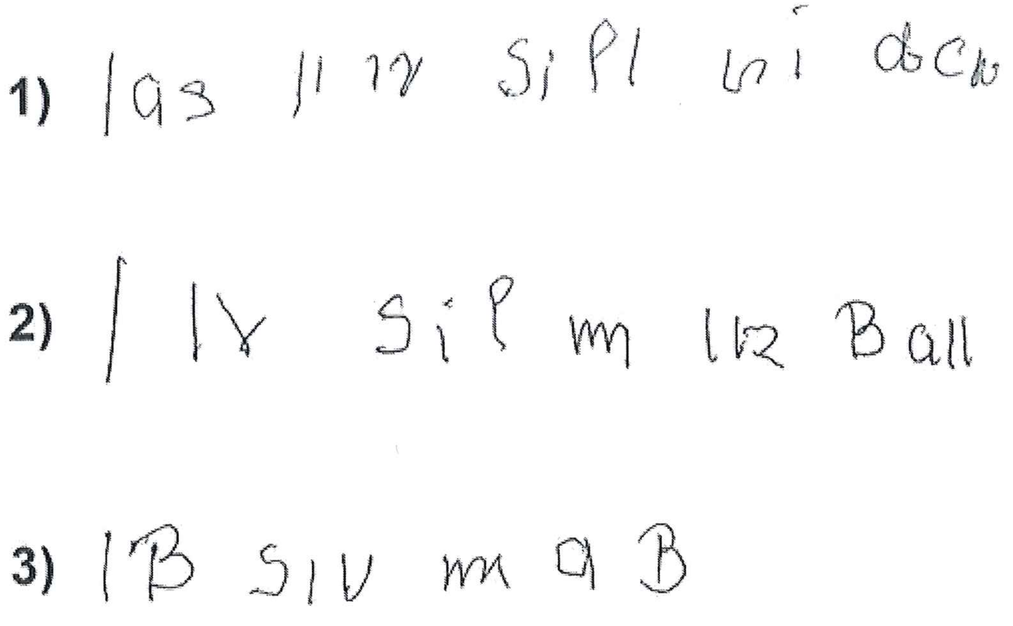

Figure 2. Agraphia. Performance on the task to write the sentence "Das Kind spielt mit dem Ball" (German for "the child plays with the ball") on 3 different occasions (1-3). The patient had the written sentence lying on the table in front of her.

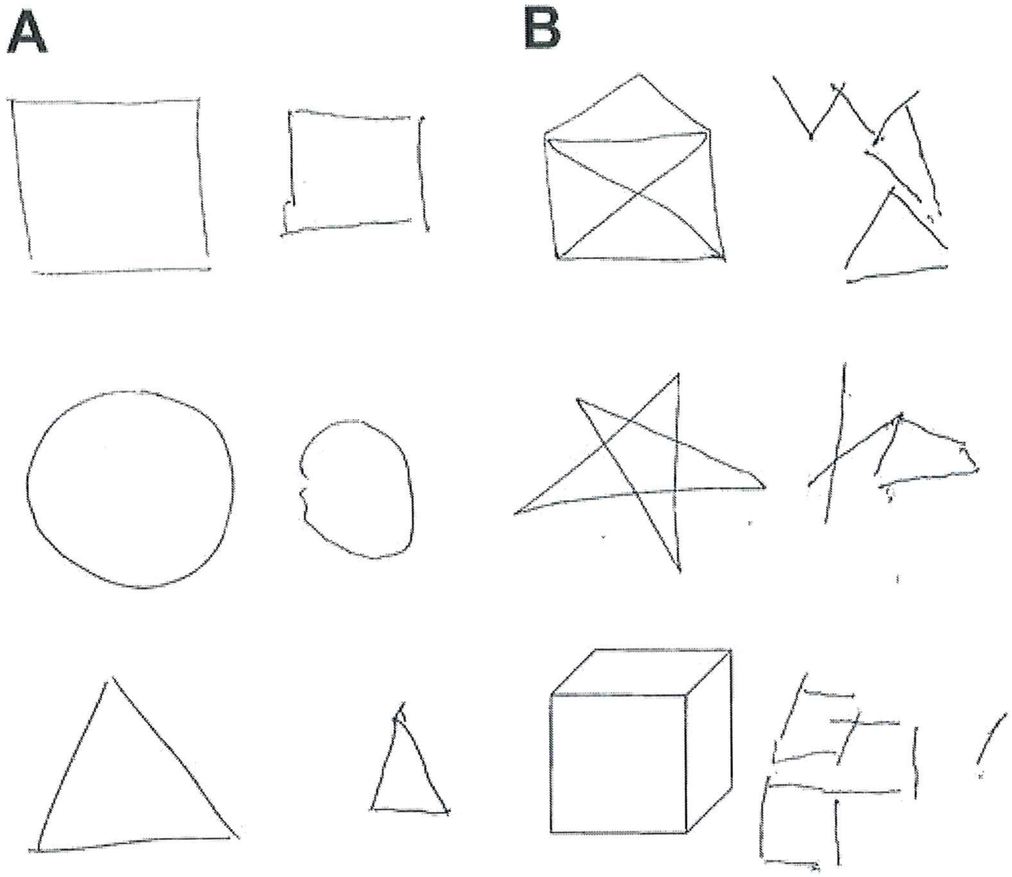

Figure 3. Visuoconstructional impairment. While drawing basic geometric figures (A) was still possible, the patient had difficulties drawing more complex figures (B), hinting to a visuoconstructional impairment.

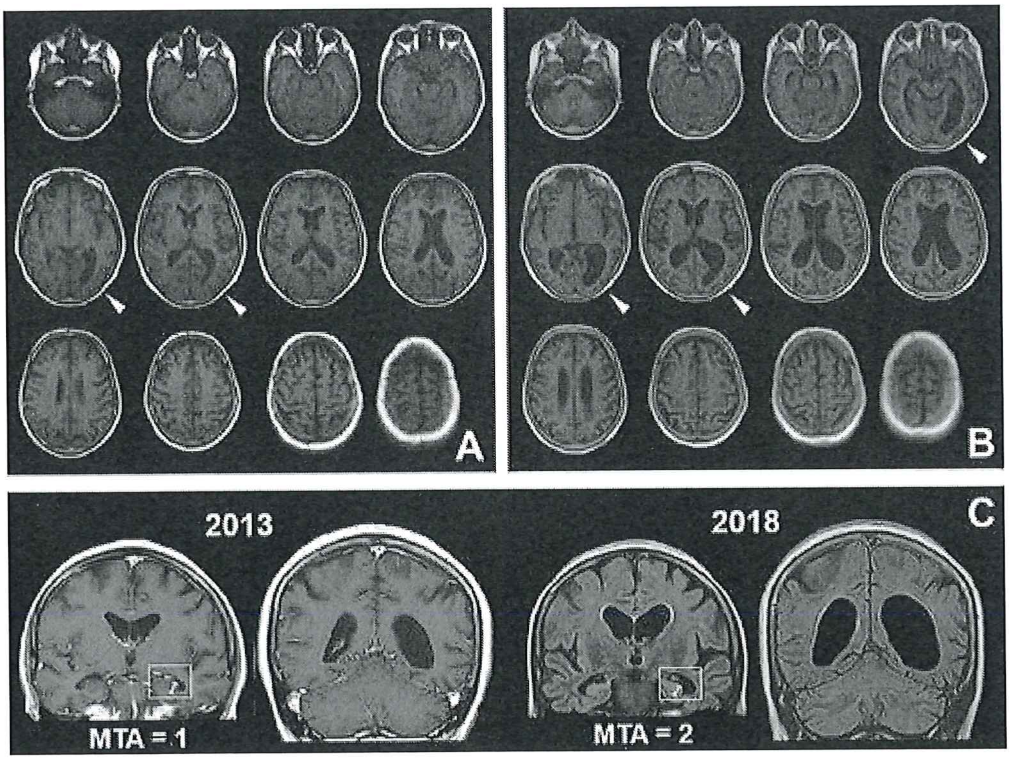

Figure 4. Brain MRI scans with progressive left parieto-occipital atrophy. In the first brain MRI scans 5 years ago (A), a dilatation of the posterior horn of the left lateral ventricle, and discrete atrophy of the surrounding cortical area (white arrows) was detected. In the recent MRI images (B), a marked ballooning of the ventricular system was found, which again appears to be most pronounced in the left posterior horn. The left parieto-occipital cortices now are shrunken immensely (white arrows). Also the coronal view (C) depicts the pronounced dilatation of the left posterior horn with only slight medial temporal atrophy due to widening of the choroidal fissure (medial temporal atrophy score [MTA] = 1) in 2013, whereas in 2018 a

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Case Reports

Full Access

Methylphenidate Treatment for Patients With Posterior Cortical Atrophy

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View Article

Posterior cortical atrophy (PCA) is the progressive and disproportionate loss of visual functions associated with neurodegeneration of posterior cortical regions.[1,2] Recent consensus criteria for PCA emphasize visuospatial and visuoperceptual deficits, relative sparing of memory and other cognitive functions, and posterior cortical abnormalities on neuroimaging.[2] The most common cause of PCA is an early-onset visual variant of Alzheimer's disease.[3] Although there is no specific treatment for PCA or clear symptom response to Alzheimer's medications, psychostimulants such as methylphenidate potentially aid in improved shifting spatial attention and, hence, overall functioni among patients.[4] Additionally, methylphenidate might improve the ability of patients with PCA to overcome lack of motivation and withdrawal consequential to visuospatial limitations. Here, case reports of three patients with PCA whose symptoms improved with methylphenidate treatment are presented.

PDF
Help

Patient 1

A 56-year-old woman presented with 2 years of progressive visual localization difficulty, particularly affecting her ability to dress herself and to drive. On dressing, she could not pick out her clothes or put her arms through the sleeves. On driving, she could not stay several years she began spending significantly more time with her children, exercising, and socializing with friends.

Patient 2

A 57-year-old woman presented with 4 years of progressive visuospatial difficulties, such as localizing nobs and finding words on pages. She demonstrated an inability to use controls or a keyboard, to find a signature block on paperwork, and to read more than two lines on a page before the "words disappear." Her MMSE score was 13/30. Her language was intact, but her memory was mild to moderately impaired. The main neurological deficits were observed on visuospatial tasks, including drawings and visual search localization. No further deficits were observed on other tasks. MRI results suggested a diagnosis of PCA, and temporoparietal and occipital hypometabolism was found on FDG-PET (Figure 1B). CSF biomarkers were consistent with Alzheimer's, and the patient was treated with donepezil (10 mg/day), memantine (10 mg/twice daily), and escitalopram (10 mg/day) for depression with anxiety.

At the 2-year follow-up, the patient's mood was generally euthymic, but she had become inactive and disengaged, primarily due to Bálint's syndrome and visuospatial constraints. She had continued treatment with donepezil and memantine, without definite treatment response. However, she was euthymic, with minimal reported anxiety while receiving escitalopram. She was started on methylphenidate (5 mg/twice daily). Although neither her visuospatial deficits nor examination results improved, her family described her as initiating more activities, as well as walking more. Additionally, and to the surprise of her family, she was able to perform more than one task at a time.

Patient 3

A 62-year-old woman presented with 1 year of progressive visuospatial difficulties localizing keypad buttons on telephones and items on desks. She also showed environmental disorientation and was unable to drive. She described difficulty voting due to an inability to locate the circles on the punch ballot. On examination, the patient's MMSE score was 27/30, her language was intact, and her memory was mildly impaired. However, she could not perform simple drawings, interpret overlapping or crosshatched figures, or localize dots or letters. Her neurological examination was otherwise normal. MRI results were nondiagnostic, but an FDG-PET showed parietal hypometabolism (Figure 1C). An amyloid PET scan was positive for amyloid neuritic plaques, consistent with PCA due to Alzheimer's disease.[6] Treatment included donepezil

PDF
Help

Diagnosing dementia

This Help Sheet provides information about the early signs of dementia, the techniques used to diagnose dementia and the importance of an early and correct diagnosis.

What are the early signs of dementia?

Because the diseases that cause dementia develop gradually, the early signs may be very subtle and not immediately obvious. Early symptoms also depend on the type of dementia and vary a great deal from person to person.

Common early symptoms include:

- Memory problems, particularly remembering recent events
- Increasing confusion
- Reduced concentration
- Personality or behaviour changes
- Apathy and withdrawal or depression
- Loss of ability to do everyday tasks

Sometimes people fail to recognise that these symptoms indicate that something is wrong. They may mistakenly assume that such behaviour is a normal part of ageing, or symptoms may develop so gradually they go unnoticed for a long time. Sometimes people may be reluctant to act even when they know something is wrong.

For the person experiencing the symptoms, the very nature of these changes within the brain may mean that the person is unable to recognise that there are changes.

Warning signs

This is a checklist of common symptoms of dementia. Go through the list and tick any symptoms that are present. If there are several ticks, consult a doctor for a complete assessment.

☐ **Memory loss that affects day-to-day function**

It's normal to occasionally forget appointments and remember them later. A person with dementia may forget things more often or not remember them at all.

☒ **Difficulty performing familiar tasks**

People can get distracted and they may forget to serve part of a meal. A person with dementia may have trouble with all the steps involved in preparing a meal.

☒ **Disorientation to time and place**

A person with dementia may have difficulty finding their way to a familiar place, or feel confused about where they are, or think they are back in some past time of their life.

☒ **Problems with language**

Everyone has trouble finding the right word sometimes, but a person with dementia may forget simple words or substitute inappropriate words, making them difficult to understand. They might also have trouble understanding others.

☒ **Problems with abstract thinking**

Managing finances can be difficult for anyone, but a person with dementia may have trouble knowing what the numbers mean or what to do with them.

☐ **Poor or decreased judgement**

Many activities require good judgement. When this ability is affected by dementia, the person may have difficulty making appropriate decisions, such as what to wear in cold weather.

☒ **Problems with spatial skills**

A person with dementia may have difficulty judging distance or direction when driving a car.

☐ **Problems misplacing things**

Anyone can temporarily misplace a wallet or keys. A person with dementia may often put things in inappropriate places.

☐ **Changes in mood, personality or behaviour**

Everyone becomes sad or moody from time to time. Someone with dementia can exhibit rapid mood swings for no apparent reason. They can become confused, suspicious or withdrawn. Some can become disinhibited or more outgoing.

☒ **A loss of initiative**

It's normal to tire of some activities. However, dementia may cause a person to lose interest in previously enjoyed activities.

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LIVING WITH POSTERIOR CORTICAL ATROPHY

By Liz, Ireland, February 24, 2016

My name is Liz Cunningham, I have lived Belfast all my life. I have been married 35 years and have two girls in their 30s getting married this year. I was diagnosed by a psychiatrist who had been treating me. She had sent me for 2 previous scans which showed nothing. She then sent me for a Pet CT Scan which showed that I had Alzheimer's, I was 47. When I was then passed over to the Dementia Elderly Services the Consultant sent me for a 3D Scan, this showed I had 3 parts of my brain affected, back, left side and frontal left lobe. I was told I had Posterior Cortical Atrophy.

Alzheimer's is a brain disease that causes a slow decline in memory, thinking and reasoning skills. The most common symptoms are consistent with damage to the posterior cortex of the brain, which is responsible for processing visual information. It is progressive and terminal, the brain tissue shrinks which causes loss of cells. The difference between Alzheimer's and PCA is that language and memory deteriorates more slowly. I have been told I'm at level 6-7 so I have already been prepared for palliative care. It affects my skills such as literacy with recalling the exact spelling of words or using a word in the right context. Following text when reading, loosing a word, sentence or rows of text and writing and typing. Numeracy is affected, as mental arithmetic is out of the question, I have major problems with money so I try to only use my card. Making movements or gestures is Awkward, and doing most household tasks takes forever to do and I still have days that disasters happen. Visual Perception causes problems trying to shower, using the correct shower gel instead of conditioner, and dressing myself at times cannot be done without help, e.g how to put my bra or pants on or buttoning a shirt. The most difficult is having visual problems, the brain does not send the signals to my eyes.

When did you first notice the effects of this disease? I was an I.T Teacher and I would have assessed people for adaptive technology. I began to get very disoriented going from my office to other offices or to the toilet. I had to ask someone how to get to my office, they thought I was joking. I had to to write substantial reports throughout the week. I couldn't make a sentence or spell words correctly or find a word to use in the right context. I became emotionally distraught. I found it difficult to teach as I could not follow instructions and forgot how to teach the software we were using. Basically I was sitting at my computer just looking at the screen most of the day. Having meetings was a nightmare

ABC NEWS

Family frustrated over NDIS as they grapple with caring for father with rare form of dementia

By Liz Hobday

Posted Mon 14 May 2018 at 6:40am, updated Mon 14 May 2018 at 6:46am

Caroline Boileau says she is struggling to get support from the NDIS for her husband.
(ABC News: Liz Hobday)

Caroline Boileau remembers her husband, Nick Ellwood, as the funniest man she's ever met.

"The first time we met, we laughed for hours, and we pretty much haven't stopped," she said.

"So even now we laugh. You have to."

Mr Ellwood was 51 years old when he was diagnosed with posterior cortical atrophy, a rare form of dementia, in 2011.

Over the next seven years the disease would take almost everything — his sight, the power of speech, even his ability to move.

Ms Boileau, a specialist nurse, now administers dozens of medications for her husband and manages every aspect of his care.

> "We feed Nick through a feeding tube in his stomach, called a peg, we wash him head to toe, we have to do his mouthwash every hour because he can't eat or drink," she said.

"We have to brush his teeth, we have to shave him, we have to change his underwear."

UP NEXT: Unexploded bomb detonated off Lord Howe Island

A woman has been arrested after a man was stabbed in Melbourne CBD

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But Wendy, now 63, can no longer read, or write. Stringing words together is a challenge.

Diagnosed with a rare form of early-onset dementia (posterior cortical atrophy) five years ago, she recently moved to an aged-care facility in Yarra Junction, east of Melbourne, when her daily needs became too much for her husband Terry.

Terry Barnett with wife Wendy Barnett, daughter Victoria Soffer and her son Leo. LUIS ENRIQUE ASCUI

As the Australian population ages and we live for longer, the number of people with dementia is predicted to soar. Last year, 237 new cases were diagnosed each day.

And the national cost will more than double in almost 40 years, from more than $14 billion this year to $37 billion by 2056, according to new research on the economic cost of dementia from Alzheimer's Australia.

But the individual costs - financial, social and familial - are also great, particularly for the families of those whose dementia is diagnosed earlier in life.

When Wendy moved into the home last November, Terry paid a $200,000 bond. He will pay $60,000 per year in out-of-pocket expenses. He also pays for extra carers to spend time with Wendy because she needs different stimulation to her fellow residents, whose average age is over 80.

Terry couldn't sell their home (now in Healesville): he still needs to live in it. They were too young for the pension. So he became his wife's full-time carer and lives on his self-managed super fund. They both had good incomes while they were working.

09/26/2014

July of 2012 I am 59 years old. I was told I have PCA a Visual Variant of Alzheimers. That I have between 8 to 12 years to live. Unlike regular Alzheimer's, the progress is much quicker, and mobility, balance and vison are affected to the point I should seek help from Services for the Blind.

The news is devastating. There is no cure, there are meds for depression and some for Alzheimer's, but that is it. I am not a shell of a man, who used to be a Military policeman, then a Special Education Teacher, and a Caregiver for the severely disabled (because teaching doesn't pay enough). I have learned a lot about 'coping' and this is one more thing to cope with. I see 4 of everything within 8 feet. I haven't seen my face in 4 years and I cannot recognize family members. They don't want to visit me. My wife is helpful, but it's all too much for her and she needs to work.

I have to re-teach myself basic skills every week as I digress. Typing is with the help of a narrator program that calls out the keystrokes and reads it back to me. This has taken me about three hours to write. I don't get to choose what works and doesn't work. The back part of my brain is dying. I am about Stage 5 out of 7 stages of PCA and I'm told that Stage 7 slips into regular Alzheimer's and when I'm bedridden I will have about a week.

COMMENTS

John Fisher

Posted: 02 Jul 2013

I would like to know if there are any facilities currently taking PCA-diagnosed patients for observation and study of this disease. My wife is now 61 and has been diagnosed with PCA. We started seeing symptoms about two years ago but the disease has progressed rapidly. She currently struggles to pick up any object, and can't handle any type of stairs without holding on to a person or fear of falling. She paces almost constantly. I have tried to document her rapid changes for doctors in Chapel Hill. She was formerly a registered nurse with great talent for painting, sewing, decorating, etc. She has never been a smoker or alcohol user and has no known hereditary issues. Her request in the past had always been to use her body for science and research should there be an opportunity. She is now being recommended for placement in a memory care facility and I see this as opportunity should there be any place looking to research this disease. As her legal guardian and power of attorney, I would welcome the opportunity to discuss any possibilities that would help fulfill her requests.

AA HH

Posted: 09 Aug 2013

My mother is 10 years into this disease. I knew she was sick when she couldn't plug the telephone cord into the wall. She also had difficulty aiming the metal part into the base when putting her seat belt on. Neurologists said they couldn't find anything for three years but I knew. As we went to Los Angeles, the neurologists there had had three cases already and they knew right away.

My mom was a doctor. She could still discuss complex topics. Her depth perception and visual fields were definitely affected. I had to take away her license. The doctors never suggested this and other family members were opposed to it initially. I knew she could have run over someone. She was scared to use stairs and had difficulty walking in general. She couldn't walk in a given direction and needed to be guided.

Eventually she began forgetting words and describing objects so that we could name them for her. Now she greatly struggles to swallow and is wheelchair bound and uncomfortable. Insurance for getting wheelchair and other goods has been a incredibly difficult with a rare diagnosis. She doesn't speak anymore. The occasional word or laugh comes out so that you know she understands the conversation. At other times, she seems completely gone. Sometimes she recognizes me sometimes she doesn't. When she was in the ER, I didn't think she recognized me but the nurse said that she was much more calm with me and my sister in the room.

Nothing can prepare you for how difficult this disease is. Some friends don't seem to believe my assessment of her and minimize it. My mom is still able to smell and taste....the only joys left. If I put a gardenia, lavender, scratch a lemon peel under her nose, she is in heaven. A hug and touch